

# U.X.L Encyclopedia of DISEASES AND DISORDERS

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VOLUME 1: A-Cl VOLUME 2: Co-G VOLUME 3: H-L VOLUME 4: M-Se VOLUME 5: Sh-Z

Rebecca J. Frey Larry I. Lutwick, Editor

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## U•X•L Encyclopedia of Diseases and Disorders

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#### Infectious

Bacteria Anthrax Chlamydia

Gonorrhea Necrotizing fasciitis Periodontal disease Plague Rheumatic fever Scarlet fever Tetanus Tooth decay Toxic shock syndrome Tuberculosis Urinary tract infection Whooping cough Virus AIDS Avian influenza Chickenpox Cold sore Common cold Ebola and Marburg hemorrhagic fevers Encephalitis Genital herpes Hand-foot-and-mouth disease Hantavirus infection Hepatitis A Hepatitis B Hepatitis C

HPV infection Infectious mononucleosis Influenza Measles Lyme disease Polio Rabies Rubella Severe acute respiratory syndrome Smallpox Staph infection Strep throat Syphilis Warts West Nile virus infection Bacteria and/or Virus Ear infection Meningitis Tonsillitis Parasite Lice infestation Malaria Toxoplasmosis Injury Burns and scalds Concussion

#### List of Entries by Type of Disease

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Gastroesophageal reflux disease Gigantism Glaucoma Headache Hearing loss Heart attack Heart diseases Heart failure Hives Hydrocephalus Hypercholesterolemia Hyperopia Hypertension Hypoglycemia Hypothermia Hypothyroidism Iron-deficiency anemia Lactose intolerance Laryngitis Learning disorders Lung cancer Marijuana use Motion sickness Myopia Obesity Obsessive-compulsive disorder Osteoarthritis Osteoporosis Panic disorder Pneumonia Postpartum depression Posttraumatic stress disorder Prematurity Scoliosis Seasonal affective disorder Seizure disorder Skin cancer Sleep apnea

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Gangrene

Leukemia Lupus Lymphoma Multiple sclerosis Narcolepsy Parkinson disease Prostate cancer

- Restless legs syndrome Reye syndrome Rheumatoid arthritis Schizophrenia Sjögren syndrome Spina bifida Strabismus
- Sudden infant death syndrome Tourette syndrome Ulcerative colitis Vitiligo

# Please Read—Important Information

The U•X•L Encyclopedia of Diseases and Disorders is a medical reference product designed to inform and educate readers about a wide variety of health issues related to diseases and injuries. Cengage Gale believes the product to be comprehensive, but not necessarily definitive. It is intended to supplement, not replace, consultation with a physician or other healthcare professional. While Cengage Gale has made substantial efforts to provide information that is accurate, comprehensive, and up-to-date, Cengage Gale makes no representations or warranties of any kind, including with 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. They are also advised to seek professional diagnosis and treatment for any medical condition, and to discuss information obtained from this book with their healthcare provider.

## Preface

"Only the curious will learn and only the resolute overcome the obstacles to learning. The quest quotient has always excited me more than the intelligence quotient." - Eugene S. Wilson (1968)

Eugene "Bill" Wilson (1900–1981) was Dean of Admission at Amherst College. He was known for his sense of humor and his genuine interest in the welfare of each student. This quote attributed to him, published in *Reader's Digest* in April of 1968, summarizes education to me, that is, it is learning itself that is most exciting, not the knowledge per se that one gathers from it. It is truly the lifelong ride of learning, not the final destination, that makes us what we are.

As both a college and medical student and now as a medical educator, I have personally seen numerous, and far too many, individuals whose goal is not learning for learning sake but rather come to me, my brilliant lecturer-wife Suzanne, or any other instructor with the unfortunately all-to-common request of "what do I have to know to pass the exam?"

Would you consult a physician who only knows what he needed to pass the test? Would you hire a tax preparer that could pass the qualifying exam but knew no more? Would you allow your child to drive an automobile with only the knowledge acquired in the Driver's Education manual? The answer to all these questions should be a resounding no.

Whether or not you are using this reference text because you are thinking about, or even planning, a career in a medical field, remember that books such as this one are limited in scope. That is, the information here should be a starting place for anyone who has the true desire to

#### Preface

understand the topic. Each entry includes further references for some additional reading, and you are encouraged to follow up on the topic. Additionally, remember that textbooks provide a snapshot of the information available at that time. The half-life of truth can be short because new information appears. Currency, that is, being current, is vital in understanding issues.

As the editor of this text, I wish each of its readers success in your future plans. "Earning a living," so to speak, implies working to live. I hope that you will have the opportunity, as I do, to truly live to work. A career can more than a job, more than just a way of paying the bills. By living to work, the work is not really work in the true sense but rather it is what you do, it is part of your essence. As it is said, "Those who love their jobs do not work a day in their lives."

Remember life is all about the ride, not the destination. Those who feel that "he who dies with the most toys wins" have missed all the scenery. Keep your eyes wide open.

#### Larry I. Lutwick, editor

## Reader's Guide

The *U*\**X*\**L* Encyclopedia of Diseases and Disorders is devoted to helping younger students and general readers understand the nature of diseases and disorders of every type, including communicable diseases, genetic disorders, common conditions, and injuries.

This book is a collection of more than 200 entries on diseases, from avian flu, to cystic fibrosis, to warts. The entries start with a definition section and highlight the basic facts of the disease to explain the causes, symptoms, and treatments. Other sections give a more detailed description, talk about demographics, and discuss the future of the disease, for example, if any new treatments are under development or if the disease is becoming more or less prevalent.

The U\*X\*L Encyclopedia of Diseases and Disorders uses everyday language when possible, and explains medical terms as they arise. Terms are also defined in Words to Know sidebars within entries, and a collected Words to Know section is included in the beginning of each book. Entries are designed to instruct, challenge, and excite less-experienced students, while providing a solid foundation and reference for students already captivated by medicine.

## Essential features of U\*X\*L Encyclopedia of Diseases and Disorders

This book contains 192 main entries and 18 overview entries. Overview entries are short descriptions of a group of disorders, like learning disorders. Each overview entry points the reader to specific entries that are part of that group. All articles in the book are meant to be understandable by anyone with a curiosity about diseases. Entries are arranged alphabetically throughout the volumes. *See also* references at the end of entries alert the readers to related entries across the three-volume set that may provide additional resources or insights each topic.

A List of Entries by Disease Type section allows readers to quickly identify diseases by types, such as infectious or genetic. Each entry contains a *Words to Know* section to help students understand important or complex terms. A general compendium of these terms is also included in the book.

A *Where To Learn More* section lists helpful print material and Web sites, while a comprehensive *General Index* guides the reader to topics and terms mentioned in the book.

Photos and color illustrations are included throughout the book where they might stimulate interest or understanding.

## **Advisors and Contributors**

While compiling this volume, the editors relied on the expertise and contributions of Rebecca J. Frey, medical writer. Frey is a freelance writer and editor who has contributed to Gale/Cengage health and medical publications since 1997. A member of the American Medical Writers Association, she completed her B.A. at Mount Holyoke College and her Ph.D. at Yale University. She lives in New Haven, Connecticut.

The editor would like to thank his contacts at Gale, Kristine Krapp and Debra Kirby, for their invaluable assistance in the technical aspects of putting this collection together. Personally, he is indebted to his wife Suzanne for her love and encouragement during this project and to their children Rachel, Zachary, Arielle, and Nina for setting such examples of how to pursue goals in life based on happiness potential, not financial reward. To their grandchildren Talora and Zev, they hope that the information in this collection serves to spark intellectual curiosity so you two will follow your hearts and minds in making this a better world.

> Larry I. Lutwick, editor Brooklyn, New York December 2008

Larry I. Lutwick MD is an academically trained Infectious Diseases physician who is Director of the Infectious Disease Section at the Brooklyn Campus of the Veterans Affairs New York Harbor Health Care System. He is also Professor of Medicine at the State University of New York, Downstate Medical School, also in Brooklyn, New York. Dr. Lutwick has authored or coauthored more than 100 scientific papers, 26 book chapters, and is the editor of several books, including (along with his wife Suzanne) *Beyond Anthrax: The Weaponization of Infectious Diseases.* He is also the creator of the medical educational tool, "Bug of the Month," in which the protagonist, a politically incorrect Infectious Disease physician named Dr. Schmeckman, solves medical mysteries.

## Words to Know

# A

**Abscess:** A collection of pus that has formed in a body cavity or hollow.

- **Abstinence:** 1.) Complete stopping of alcohol consumption. 2.) Not having sexual intercourse with anyone.
- **Acclimation:** The process of adjusting to seasonal climate changes or to a new climate.
- **Accommodation:** The medical term for the eye's ability to change its focus automatically for viewing objects at different distances.
- **Acetaldehyde:** A colorless liquid chemical that is produced when the body begins to digest alcohol. A chemical that causes hangovers after heavy drinking, it also contributes to fetal alcohol syndrome.
- Achilles tendon: The tendon that connects the calf muscle to the back of the heel. Tendinitis in the Achilles tendon is common in sports that involve running and jumping.
- **Acromegaly:** A condition in which a person's body produces too much growth hormone in adult life.
- Actinic keratosis: A patch of thickened or scaly skin caused by sun exposure. It is not itself a form of skin cancer but may develop progressively into a skin cancer.
- **Acupuncture:** A form of alternative medicine in which very fine needles are inserted into the skin at specific points on the body for pain relief.

- Acute: Referring to a disease or symptom that is severe or quickly worsens.
- **Addiction:** A chronic disease characterized by compulsive drug use and by long-lasting chemical changes in the brain.
- Adipose tissue: Fatty tissue.
- Adrenaline: A hormone that can be used in medicine to open the breathing passages in patients with severe tissue swelling. It is also called epinephrine.
- Affective disorder: A type of mental disorder characterized by disturbed emotions and feelings rather than problems with memory, thinking, or learning.
- **After drop:** A term that doctors use to refer to lowering of the body's core temperature that continues while the person is being rewarmed.
- **Against-the-rule astigmatism:** A type of astigmatism in which the eye sees horizontal lines more clearly than vertical lines.
- **Agoraphobia:** An irrational fear of venturing outside the home or into open spaces, so strong that a large number of activities outside the home are limited or avoided altogether. Agoraphobia is often associated with panic attacks.
- **Allergen:** A substance that causes an allergic reaction in individuals who are sensitive to it.
- **Alveoli (singular, alveolus):** Tiny air sacs in the lungs where carbon dioxide in the blood is exchanged for oxygen from the air.
- Amaranth: An herb that produces seeds used as grain in India, Nepal, Mexico, and parts of South America.
- **Amblyopia:** Dimness of sight in one eye without any change in the structure of the eye. It is also known as lazy eye.
- Amenorrhea: Stopping of normal menstrual periods.
- **Amino acids:** A group of twenty compounds that are the building blocks of proteins in humans and other animals.
- **Ammonia:** A chemical produced during the breakdown of protein in the body. It is usually converted in the liver to another chemical called urea and then discharged from the body in the urine.
- **Amputation:** Surgical removal of a limb.

- **Anabolic:** Referring to tissue building. Anabolic steroids build up muscle and bone tissue.
- Anaerobic: Capable of living in the absence of oxygen.
- **Anaphylaxis:** A severe allergic reaction to a trigger (most commonly a food, medication, insect sting, or latex) that involves most major body systems.
- **Androgen:** The generic term for the group of male sex hormones produced by the body.
- **Anemia:** A condition in which a person's blood does not have enough volume, enough red blood cells, or enough hemoglobin in the cells to keep body tissues supplied with oxygen.
- **Aneurysm:** A weak or thin spot on the wall of an artery.
- Angina: Chest pain caused by an inadequate supply of blood to the heart muscle.
- **Angioedema:** The medical term for the swelling of tissues can be part of an allergic reaction.
- **Antibody:** A protein found in blood that is specific to a particular foreign substance, which may be an allergen or a disease organism. The antibody identifies that antigen and neutralizes it.
- **Anticipation:** A condition in which the symptoms of a genetic disorder appear earlier and earlier in each successive generation.
- **Antidepressant:** A type of drug given to treat eating disorders as well as mood disorders like anxiety and depression.
- **Antiemetic:** A type of drug given to control nausea and vomiting.
- **Anti-psychotics:** A group of drugs used to treat schizophrenia. The older anti-psychotic drugs are also called neuroleptics.
- **Antispasmodic:** A type of drug given to relieve the cramping of the intestines or other muscles.
- **Aorta:** The large artery that carries blood away from the heart to be distributed to the rest of the body.
- **Aortic dissection:** A tear in the wall of the aorta that allows blood to seep between the layers of tissue that form the artery and push the layers apart.
- **Aphthous ulcer:** The medical term for canker sore.
- **Apnea:** Temporary stopping of breathing.

**Appendectomy:** Surgical removal of the appendix.

Arboviruses: A family of viruses spread by blood-sucking insects.

**Area postrema:** The part of the brain stem that controls vomiting.

- **Arson:** The intentional setting of a fire in a building or other property. Arson is a criminal act in the United States.
- **Arthroscopy:** The use of a small device called an arthroscope to look inside and diagnose or treat an arthritic joint.
- **Aseptic meningitis:** A term that is sometimes used for meningitis that is not caused by bacteria.

Aspie: An informal name for a person with Asperger syndrome.

- **Aspiration:** The entry of food, liquids, or other foreign substances into the lungs during the breathing process.
- **Astigmatism:** A vision problem caused by irregularities in the shape of the cornea or the lens of the eye.
- Asymptomatic: Having no symptoms.
- **Atherosclerosis:** Stiffening or hardening of the arteries caused by the formation of plaques within the arteries.
- **Atopic disease:** Any allergic disease that affects parts of the body that are not in direct contact with the allergen. Asthma, eczema, and hay fever are all atopic diseases.
- **Atopy:** The medical term for an allergic hypersensitivity that affects parts of the body that are not in direct contact with an allergen. Hay fever, eczema, and asthma are all atopic diseases.
- Atrium (plural, atria): One of the two upper chambers of the heart.
- Audiologist: A health care professional who is specially trained to evaluate hearing disorders.
- **Auditory:** Pertaining to the sense of hearing.
- Aura: A symptom that precedes migraine headaches in some people. The person may see flashing or zigzag lights, or have other visual disturbances.
- **Autism:** A developmental disorder that appears by three years of age and is characterized by limited communication skills, difficulties in communicating with others, and difficulties forming relationships.
- **Autoantibody:** An antibody formed in reaction against the tissues of the individual producing it.

- Autoimmune disease: A disease in which the body's immune system attacks its own cells and tissues.
- Autoimmune disorder: A disorder characterized or caused by autoantibodies that attack the cells or organs of the organism producing them.
- Automatic behavior: Activity that a person with narcolepsy can carry out while partially awake but is not conscious of at the time and cannot recall afterward.
- **Autopsy:** The examination of a body after death to determine the cause of death.

Avian: Pertaining to birds.

## В

- **B cell:** A type of white blood cell produced in the bone marrow that makes antibodies against viruses.
- **Babesiosis:** A malaria-like disease that can be transmitted by ticks.
- **Baby blues:** An informal term for the temporary sad feelings some mothers feel for a week or so after childbirth. It is less serious than postpartum depression and usually goes away by itself.
- Bacteremia: The presence of bacteria in the bloodstream.
- **Bedsore:** A type of wet gangrene that develops when a bedridden person cannot turn over to relieve pressure on soft tissue caused by the weight of the body. Bedsores are sometimes called pressure ulcers.
- Benign: Not cancerous.
- **Benign prostatic hypertrophy (BPH):** A noncancerous condition in which the swelling of the prostate gland squeezes the urethra and causes difficulty in urination.
- **Beta blockers:** A group of drugs given to treat abnormal heart rhythms and reduce the risk of aortic dilation in MFS patients.
- **Bile:** A yellow-green fluid secreted by the liver that aids in the digestion of fats.
- **Binge:** An episode of eating in which a person consumes a larger amount of food within a limited period of time than most people would eat in similar circumstances.

- **Binge drinking:** A period of heavy drinking that lasts for two days or longer.
- **Biofeedback:** An alternative treatment for headaches (and other conditions) that consists of teaching patients to consciously control their blood pressure, muscle tension, temperature, and other body processes.
- **Biological therapy:** An approach to cancer treatment that is intended to strengthen the patient's own immune system rather than attack the cancer cells directly.
- **Biomarker:** A substance produced by the body that is distinctive to a particular disease and can be used to identify its presence or track its progress.
- **Bioterrorism:** The use of disease agents to frighten or attack civilians.
- **Biphasic reaction:** A recurrence of the symptoms of anaphylaxis about six to eight hours after the first episode.
- Blackout: Alcohol-related memory loss.
- **Bladder:** A hollow organ in the lower abdomen that collects urine from the kidneys and stores it prior to urination.
- **Blunt:** A cigar that has been cut open and refilled with marijuana.
- **Body mass index:** BMI. An indirect measurement of the amount of body fat. The BMI of adults is calculated in English measurements by multiplying a person's weight in pounds by 703.1, and dividing that number by the person's height in inches squared.
- **Bone marrow:** The soft spongy tissue inside the long bones of the body where blood cells are formed.
- **Bong:** A water pipe used to smoke marijuana.
- **Botulism:** A rare but potentially fatal paralytic illness caused by a bacterial toxin in contaminated food.
- **Brain stem:** The lowest part of the brain that connects directly to the spinal cord. It controls such basic life functions as breathing, blood pressure, and heart beat.
- **Bronchiole:** A very small thin-walled air passage in the lungs that branches off from a bronchus.
- **Bronchodilator:** A type of drug that opens up the bronchi, increasing airflow and relieving wheezing and other asthma symptoms.

- **Bronchoscope:** A flexible lighted tube that can be inserted into the passages leading to the lungs for examination or treatment.
- **Bronchus (plural, bronchi):** One of the two major divisions of the airway that lead into the right and left lungs.
- **Bubo:** A swollen lymph node in the neck, armpit, or groin area.

C

- **Café-au-lait spots:** Brownish-white birthmarks that appear as part of a nervous system disorder that can cause scoliosis in some children. Café au lait is the French expression for "coffee with milk" and describes the color of the spots.
- **Capsule:** The outermost layer of the lens of the eye.
- **Carcinoma:** The medical term for any type of cancer that arises from the skin or from the tissues that line body cavities.
- **Carcinoma in situ:** A cancer that has not spread or is still in one location in the body.
- **Cardiac arrest:** Heart attack; a condition in which the circulation of the blood stops abruptly because the heart stops beating.
- Carditis: Inflammation of the heart.
- Caries: The medical name for tooth cavities.
- **Carrier:** A person who is infected with a disease and can spread it to others but who has no symptoms of the disease.
- **Cartilage:** A type of dense connective tissue that serves to cushion bones within joints.
- **Case management:** An approach to healthcare based on personalized services to patients.
- **Cataplexy:** Sudden loss of tone in the voluntary muscles.
- **Catatonia:** A condition in which a person sits motionless for long periods of time and does not respond to others.
- **Catheter:** A thin tube inserted into the urethra to drain urine from the bladder.
- **Cerebellum:** The part of the brain at the lower back of the head just above the brain stem.

- **Cerebral cortex:** The part of the brain that controls thinking, memory, paying attention, decision making, and using language.
- **Cervix:** The neck or lowermost part of a woman's uterus that opens into the vagina.
- **Chancre:** A painless ulcer that forms on the skin during the early stage of syphilis.
- **Chelation therapy:** A form of treatment to reduce overly high levels of iron (or other metals) in the body by giving the patient a chemical that allows the body to get rid of the excess metal in urine or stool.
- **Chiropractic:** A form of alternative medicine that treats disorders of the joints and muscles by adjusting the patient's spine or other joints.
- **Chlamydia:** A sexually transmitted disease caused by a bacterium that is a common cause of eye infections.
- **Cholesterol:** A fatty substance produced naturally by the body that is found in the membranes of all body cells and is carried by the blood.
- **Chorea:** A general term for movement disorders marked by loss of coordination and involuntary motions of the head and limbs.
- **Chorionic villus sampling (CVS):** A prenatal test that involves taking a small sample of the placenta, the organ that forms inside the uterus during pregnancy and supplies the baby with oxygen and nutrients carried by the blood.
- **Chronic:** Referring to a disease or symptom that goes on for a long time, tends to recur, and usually gets worse slowly.
- **Circadian rhythm:** The medical name for the daily sleep/wake cycle in humans.
- **Cirrhosis:** Disruption of normal liver function by the formation of scar tissue and nodules in the liver. It is most commonly caused by alcoholism or hepatitis C.
- **Clap:** A slang term for gonorrhea.
- **Clinically isolated syndrome (CIS):** A term applied to patients who have had one episode of illness that suggests they have a disease but do not yet meet the full criteria for diagnosis.

- **Closed-head injury:** An injury to the head in which the skull is not broken or penetrated.
- Clubbing: Thickening of the tips of the fingers or toes.
- **Coagulation cascade:** The complex process in which platelets, coagulation factors, and other chemicals in the blood interact to form a clot when a blood vessel is injured.
- **Coagulation factors:** Proteins in blood plasma involved in the chain of chemical reactions leading to the formation of blood clots. They are also called clotting factors.
- Cochlea: A snail-shaped fluid-filled chamber in the inner ear.
- **Cognitive:** Related to thinking, memory, and other conscious intellectual activities or processes.
- **Cognitive-behavioral therapy (CBT):** An approach to therapy that aims at changing distorted thinking patterns, beliefs, and behaviors in order to change the patient's feelings.
- **Colon:** The part of the large intestine that extends from the cecum to the rectum.
- **Colonization:** The process by which bacteria form colonies in or on the bodies of humans and other animals.
- **Comedo (plural, comedones):** The medical term for a whitehead or blackhead.
- **Community-acquired:** Referring to a disease that a person gets in the course of ordinary activities rather than in a hospital or clinic.
- **Compression fracture:** A fracture caused by the collapse of a vertebra in the spinal column, usually caused either by trauma or by weakening of the bone in osteoporosis.
- **Compulsion:** A repeated behavior or mental act carried out to control or neutralize obsessions.
- **Conditioning:** The process of becoming physically fit through a program of diet, exercise, and rest.
- Congenital: Present at birth.
- **Congenital rubella syndrome (CRS):** A group of birth defects that may affect a baby born to a mother who had rubella during the first three months of pregnancy.

- **Conjunctiva (plural, conjunctivae):** The clear membrane that covers the white part of the eyeball and lines the eyelids.
- **Contact dermatitis:** Inflammation of the skin caused by direct contact with an allergen or irritating substance, such as poison ivy, certain dyes, or certain metals.
- **Contracture:** Shortening or tightening of the muscles surrounding certain joints that limits the movement of the joints.
- **Cooley's anemia:** Another name for the most severe form of beta-thalassemia.
- **Coprolalia:** The medical term for uncontrollable cursing or use of dirty words.
- **Cornea:** The transparent front part of the eye where light enters the eye.
- **Cortex:** The part of the lens underneath the capsule.
- **Cortisol:** A hormone produced by the adrenal glands near the kidneys in response to stress.
- **Co-sleeping:** Allowing a baby to sleep in the same bed as its parents. It is also called bed sharing.
- **Crabs:** A slang term for pubic lice.
- Cretinism: A form of hypothyroidism found in some newborns.
- **Cryotherapy:** The use of extreme cold to destroy cancerous tumors or other diseased tissue. It is also called cryosurgery.
- **Cushing syndrome:** A disorder caused by the excess secretion of cortisol by the pituitary gland.
- Cutaneous: Pertaining to the skin.
- **Cyanosis:** A blue discoloration of the lips, inside of the mouth, and nail beds caused by lack of oxygen in the blood vessels near the skin surface.
- **Cycling:** Using steroids in periods of several weeks or months (a time cycle) separated by short rest phases of not using the drugs.
- Cyclothymia: A mild form of bipolar disorder.

**Cyst:** A capsule or sac containing a parasite in its resting stage.

Cystitis: The medical term for an infection of the urinary bladder.

# D

- **Dander:** Tiny skin, feather, or fur particles from household pets that cause allergic reactions in some people.
- **Debridement:** The medical term for the surgical removal of dead or damaged soft tissue.
- **Decibel (dB):** A unit of measurement for expressing the relative intensity of sounds.
- **Decoding:** In education, the ability to associate letters of the alphabet with sounds.
- **Degenerative disorder:** A type of disorder in which a person gradually loses certain abilities that he or she had acquired at an earlier age.
- **Dehydration:** Loss of water from the body. It may be caused by fever, vomiting, diarrhea, or excessive sweating.
- **Delirium:** A suddenly developing mental disturbance characterized by confused thinking, difficulty focusing attention, and disorientation.
- **Delirium tremens:** A severe physical reaction to withdrawal from alcohol in which the person hallucinates and has unstable blood pressure and breathing patterns.
- **Delusion:** In medicine, a false belief that a person holds to despite evidence or proof that it is false.
- **Dementia:** Loss of memory and other mental functions related to thinking or problem-solving.
- **Dengue:** A tropical disease caused by a virus similar to the virus that causes West Nile infection. It is also spread by mosquitoes.
- **Dentin:** A firm tissue that lies between the enamel and the pulp of a tooth.
- **Dermabrasion:** Technique for making acne scars less noticeable by removing the top layer of skin with a rapidly rotating wire brush or a sandy material.
- Dermatitis: The medical term for inflammation of the skin.
- **Dermatographism:** A type of hives produced by scratching or stroking the skin.
- **Dermatologist:** A doctor who specializes in diagnosing and treating diseases and disorders of the skin.

- **Dermatology:** The branch of medicine that deals with skin problems and disorders.
- **Dermis:** The lower layer of skin that contains blood vessels, sweat glands, and hair follicles.
- **Desensitization:** A form of treatment for allergies that involves a series of shots containing the allergen to reduce the patient's sensitivity to that particular trigger. Desensitization is also called immunotherapy.
- **Detoxification:** A process or treatment program for clearing an alcoholic's body of alcohol. It usually includes medications to help manage the physical symptoms of withdrawal.
- **Diagnosis of exclusion:** A diagnosis that the doctor arrives at by ruling out other diseases one by one rather than making the diagnosis on the basis of laboratory tests or imaging studies, or other test results.
- **Dialysis:** A process in which the blood of a patient with kidney failure is cleansed of the body's waste products by being pumped through a machine that filters the blood and then returns it to the body.
- **Diaphragm:** A sheet of muscle extending across the bottom of the rib cage that separates the chest from the abdomen.
- **Diastolic blood pressure:** The blood pressure when the heart is resting between beats.
- **Directly observed therapy (DOT):** Treatment in which nurses or health care workers administer medications to patients in a clinic or doctor's office to make sure that the patients take the drugs correctly.
- **Disseminated gonococcal infection (DGI):** A complication of gonorrhea in which the disease organisms get into the bloodstream and cause arthritis, eye disease, skin rashes, or inflammation of the heart valves.
- **Diuretic:** A type of drug that increases the body's production of urine.
- **Dopamine:** A chemical produced in the brain that is needed to produce smooth and controlled voluntary movements.
- **Dronabinol:** A medication that contains synthetic THC, given to relieve nausea and improve appetite in AIDS and cancer patients.
- Duodenum: The first part of the small intestine.
- **Dysthymia:** A mood disorder characterized by a long-term low-key depression.
- **Dystrophin:** A protein found in muscle whose absence or defectiveness is one of the causes of muscular dystrophy.



- **Echinacea:** A plant native to the eastern United States that is thought by some to be a useful cold remedy. It is also known as purple coneflower.
- **Ectopic pregnancy:** A pregnancy in which the fertilized egg starts growing outside the uterus, usually in the abdomen or in the tubes leading to the uterus.
- **Effusion:** The medical term for an abnormal collection of fluid in a body cavity.
- **Ehrlichiosis:** A tick-borne disease found primarily in dogs that can also be transmitted to humans.
- **Electroconvulsive therapy (ECT):** A form of treatment for severe depression that consists of passing a low dose of electric current through the patient's brain under anesthesia.
- **Electrolytes:** Minerals that are essential for proper body functioning. They include potassium, sodium, calcium, and magnesium.
- **Embolus:** The medical term for a clot that forms in the heart and travels through the circulatory system to another part of the body.
- **Embryo:** The medical term for an unborn baby from the time of conception to the end of its first eight weeks of life.
- **Emerging infectious disease (EID):** A disease that has become more widespread around the world in the last twenty years and is expected to become more common in the future.
- **Enamel:** The hard, smooth, white outer surface of a tooth.
- **Encephalitis:** Inflammation of the brain.
- **Endemic:** A term applied to a disease that maintains itself in a particular area without reinforcement from outside sources of infection.
- **Endocarditis:** An inflammation of the tissues lining the inside of the heart and its valves.
- **Endocrine system:** A system of small organs located throughout the body that regulate metabolism, growth and puberty, tissue function, and mood. The thyroid gland is part of the endocrine system.
- **Endocrinologist:** A doctor who specializes in disorders of the pancreas and other glands.
- Endophthalmitis: Inflammation of the tissues inside the eyeball.

- **Epidemiology:** The branch of medicine that deals with the frequency, distribution, and control of disease in a population.
- **Epidermis:** The outermost layer of the skin.
- **Eradication:** The complete elimination of a disease.
- **Erythema chronicum migrans (EM):** The medical name for the distinctive rash that is often seen in early-stage Lyme disease.
- **Esophagus:** The muscular tube that carries food downward from the lower throat to the stomach.
- **Essential hypertension:** High blood pressure that is not caused by medications, pregnancy, or another disease.
- **Estrogen:** A female hormone produced in the ovaries.
- Euphoria: An exaggerated feeling of well-being.
- **Eustachian tube:** The passageway that connects the middle ear with the upper throat.
- Eustress: A term that is sometimes used to refer to positive stress.
- **Euthanasia:** Sometimes called mercy killing; the act of killing a hope-lessly ill human or pet in a painless way.
- **Eversion injury:** An ankle injury caused when the foot is suddenly forced to roll outward.
- **Exophthalmos:** Abnormal protrusion of the eyeballs.
- **Exotoxin:** A toxin secreted by a bacterium or other disease organism into the body tissues of an infected individual.



- **Failure to thrive:** A term used to describe children whose present weight or rate of weight gain is markedly lower than that of other children of their age and sex.
- Fascia: A sheet of connective tissue that covers and binds together the muscles, glands, blood vessels, and internal organs of the body.
- **Fasting hypoglycemia:** A type of hypoglycemia in people without diabetes that is caused by hormone deficiencies, medication side effects, or tumors rather than by reaction to a sugar-rich meal.

- **Fatal familial insomnia (FFI):** A very rare inherited disease in which the person dies of sleeplessness.
- Fatigue: A feeling of weariness or tiredness after work, exercise, or emotional stress.
- **Female athlete triad:** A group of three symptoms that often occur together in female athletes: amenorrhea, osteoporosis, and disordered eating.
- Fibro fog: A term that has been coined to describe memory loss and difficulty concentrating in fibromyalgia patients.
- **Fibroblast:** A type of cell that provides structure during the healing of a broken bone or other wound.
- Fibrosis: The medical term for the formation of scar tissue.
- Filaggrin: A protein in the skin that is defective or lacking in some patients with eczema.
- **Filovirus:** The category of viruses that includes Ebola and Marburg viruses. Filoviruses look like long pieces of thread under a microscope.
- **Fistula:** An abnormal tunnel or passage that forms between one part of the intestine and another or between the intestine and the body surface.
- **Fixation:** The medical term for holding a broken bone in its correct position to speed healing and prevent further injury.
- Flare: A return or worsening of symptoms.
- Flashback: A temporary reliving of a traumatic event.
- **Folic acid:** A form of vitamin B<sub>9</sub> that helps to prevent spina bifida.
- Follicle: Small canal in the skin surrounding the root of a hair.
- **Forchheimer spots:** Tiny reddish spots that appear inside the mouth of a patient with scarlet fever.
- **Fragility fracture:** A fracture that occurs as a result of a fall from standing height or less. A person with healthy bones would not suffer a broken bone falling from a standing position.
- **Fulminant:** Referring to any disease or condition that strikes rapidly and is severe to the point of being life-threatening.
**Fundoplication:** A surgical procedure in which the upper part of the stomach is wrapped around the lower end of the esophagus to prevent stomach acid from rising into the esophagus.

## G

Gait: A person's characteristic pattern of walking.

Gangrene: Decay and death of soft tissue due to loss of blood supply.

Gastric: Related to the stomach.

**Gastroenterologist:** A doctor who specializes in diagnosing and treating diseases of the digestive system.

**Gene therapy:** An approach to treating disease by inserting healthy genes into a person's genetic material or by inactivating defective genes.

**Genotype:** The genetic makeup of a cell or organism.

- **Germ cell:** A cell involved in reproduction. In humans the germ cells are the sperm (male) and egg (female). Unlike other cells in the body, germ cells contain only half the standard number of chromosomes.
- **Gestational:** Pertaining to pregnancy.
- **Gestational age:** An infant's age at birth counting from the date of the mother's last menstrual period.
- **Gigantism:** Excessive production of growth hormone in children who are still growing.
- Gingivitis: The medical term for inflammation of the gums.
- **Glial cells:** Cells in brain tissue that hold nerve cells in place, supply them with oxygen and nutrients, and remove dead nerve cells.

**Glioma:** A type of brain tumor that starts in the glial cells.

- **Glucagon:** A hormone secreted by the pancreas that raises blood sugar levels by signaling the liver to convert glycogen to glucose.
- **Glucometer:** A small blood testing device that can be used to screen for diabetes or used at home to monitor blood sugar levels.
- **Gluten:** A protein found in certain grains, particularly wheat, barley, and rye.
- **Glycogen:** A form of glucose that is stored in the liver as an energy reserve.

**Goiter:** A swelling in the neck caused by an enlarged thyroid gland.

**Gonococcus:** The bacterium that causes gonorrhea.

- **Gout:** A disorder of the large toe or other joints caused by deposits of uric acid crystals in the affected joint.
- **Group A streptococcus:** A sphere-shaped bacterium that grows in long chains and causes strep throat as well as scarlet fever and some forms of tonsillitis.
- **Growth plate:** A cartilage plate in the long bones of children where the lengthening of bone takes place.
- Guarding: Stiffening of the muscles in response to a doctor's touch.
- **Gumma:** A soft noncancerous growth of tissue found in patients with tertiary syphilis.

H

- **Hair cells:** Special cells in the cochlea that convert the movement of the fluid inside the cochlea into electrical signals that travel to the brain via the auditory nerve.
- Hallucination: Perceiving something that is not really there. Hallucinations can affect any of the five senses.
- **Hard palate:** A thin bony plate located in the front portion of the roof of the mouth.
- **Hashish:** A concentrated resin prepared from the flowering tops of hemp plants.
- Hashitoxicosis: A temporary phase in some patients with Hashimoto disease in which there is too much thyroid hormone in the blood due to leakage from damaged and dying cells in the thyroid gland.
- **Heat illness:** A general term for heat-related disorders, ranging from heat cramps (the mildest) to heat stroke (the most serious).
- **Heelstick:** A method for taking a sample of blood from a newborn by pricking the baby's heel with a needle and collecting a drop or two of blood on special filter paper.
- **Hematocrit:** The proportion of blood volume occupied by red blood cells.

- Hematologist: A doctor who specializes in diagnosing and treating disorders of the blood.
- **Hemoglobin:** An iron-containing protein in red blood cells that carries oxygen from the lungs to the rest of the body.
- **Hepatitis:** A general term for inflammation of the liver. It can be caused by toxic substances or alcohol as well as infections.
- Herpetiform: Resembling blisters caused by herpes.
- **Hiatal hernia:** A condition in which the upper part of the stomach bulges upward into the chest cavity through a weak spot in the diaphragm.
- **Highly active antiretroviral therapy (HAART):** An individualized combination of three or more antiretroviral drugs used to treat patients with HIV infection. It is sometimes called a drug cocktail.
- **Histamine:** A chemical contained in mast cells that is released during an allergic reaction.
- Hit: A single intake of marijuana smoke from a joint or bong.
- **Holoprosencephaly:** A disorder in which a baby's forebrain does not develop normally. The infant's brain fails to divide into two cerebral hemispheres; this failure in turn leads to facial deformities and abnormal brain structure and function.
- **Hormone:** Any chemical produced by living cells that stimulates organs or tissues in parts of the body at some distance from where it is produced.
- **Hospice:** A facility or program for meeting the spiritual as well as the physical needs of people who are terminally ill.
- Host: An organism that is infected by a virus, bacterium, or parasite.
- **Hydrocephalus:** Abnormal accumulation of cerebrospinal fluid within the cavities inside the brain.
- **Hydrops fetalis:** The most severe form of alpha thalassemia, leading to death before or shortly after birth.
- **Hyperammonemia:** Overly high levels of ammonia in the blood; it often indicates liver damage.
- **Hyperarousal:** A state of increased emotional tension and anxiety, often including jitteriness and being easily startled.

- **Hyperbaric oxygen (HBO):** Oxygen that is delivered to a patient in a special chamber at two to three times normal atmospheric pressure.
- **Hyperextension:** Stretching or moving a part of the body beyond its normal range of motion.
- Hyperopia: The medical term for farsightedness.
- **Hyperthyroidism:** A disease condition in which the thyroid gland produces too much thyroid hormone.
- **Hypnagogic:** Referring to the period of partial alertness on the boundary between sleeping and waking.
- **Hypocretin:** A protein produced by certain brain cells that promotes wakefulness and helps to regulate the sleep/wake cycle. It is also known as orexin.
- **Hypomania:** A less severe form of mania that does not interfere with normal functioning.
- **Hypothalamus:** The part of the brain that controls body temperature, hunger, thirst, and response to stress.
- **Hypothyroidism:** A disease condition in which the thyroid gland does not produce enough thyroid hormone.
- **Hypotonia:** The medical term for poor muscle tone.

- **Ideal weight:** Weight corresponding to the lowest death rate for individuals of a specific height, gender, and age.
- **Identical twins:** Twins that develop from a single fertilized egg that divides to form two separate embryos.
- Idiopathic: The medical term for a disorder whose cause is unknown.
- **Immunoglobulin E (IgE):** An antibody in blood that activates mast cells during an allergic reaction.
- **Incest:** Sexual activity between closely related persons, often within the immediate family.
- **Indolent:** The medical term for a tumor or disease that grows or develops slowly.

- **Infestation:** A condition in which a parasite develops and multiplies on the body of its host rather than inside the body.
- **Inhalation:** The part of the breathing cycle in which a person takes in air from the outside.
- **Insulin:** A hormone secreted by the pancreas that causes the cells in the liver, muscle and fatty tissues of the body to use the glucose carried in the bloodstream after a meal.
- **Intractable:** Referring to a disease or disorder that cannot be easily treated or cured.
- **Inversion injury:** A type of ankle injury caused when the foot is suddenly forced to roll inward.
- Involuntary: Not under the control of the will.
- **Iris:** The circular colored structure at the front of the eyeball that controls the amount of light entering the eye by changing the size of the pupil.
- **Irradiation:** A technique for treating raw meat and poultry with gamma rays, x rays, or electron beams to destroy disease organisms.
- **Ischemia:** Loss of blood supply to a tissue or organ resulting from the blockage of a blood vessel.

## J

- Jaundice: A yellowish discoloration of the skin and whites of the eyes caused by increased levels of bile pigments from the liver in the patient's blood.
- Jet lag: A sleep disorder or disturbance in the sleep/wake cycle related to rapid travel across time zones.
- Joint: A cigarette made with marijuana instead of tobacco.



**Karyotype:** A photomicrograph of the chromosomes in a single human cell. Making a karyotype is one way to test for genetic disorders.

- **Keratoconus:** An eye disorder in which the tissue of the cornea grows thinner over time.
- Koplik spots: Small reddish spots with white centers seen on the tissues lining the cheeks in early-stage measles.
- **Kuru:** A fatal brain disease related to CJD that was epidemic in Papua New Guinea in the mid-1950s. Kuru is thought to have been spread by cannibalism.

C

- **Lactase:** An enzyme that breaks down lactose into simpler sugars during the process of digestion.
- **Lactose:** A complex sugar found in milk and other dairy products. It is sometimes called milk sugar.
- Lactose intolerance: An inability to digest lactose, the form of sugar found in milk and milk products.
- **Laparoscope:** A fiberoptic instrument resembling a telescope that can be inserted through a small incision to allow a doctor to see the inside of the abdomen during surgery.
- Larva: The immature form of an insect.
- **Larynx:** The medical name for the voice box located at the base of the throat.
- **Latent:** Referring to a disease that is inactive.

**Lesion:** A general term for any skin injury.

- **Levothyroxine:** The chemical name for the synthetic thyroid hormone given to treat Hashimoto disease.
- Ligament: A tough fibrous band of tissue that joins bones together.

**Lobule:** One of the glands in the breast that produce milk.

- **Lymph nodes:** Part of the lymphatic system, the lymph nodes trap foreign particles and are important to defend the body from disease.
- **Lymphocyte:** A type of white blood cell that fights infection. Lymphocytes are divided into two types, T cells (produced in the thymus gland) and B cells (produced in the bone marrow).
- Lymphoma: A type of cancer that affects the lymphatic system.

## M

- **Macule:** A spot on the skin or patch that is different in color from normal skin but is usually not raised up above the skin surface.
- **Mad cow disease:** A prion disease that affects cattle and can be transmitted to humans by eating meat from infected cattle.
- **Malabsorption:** Inability to absorb the nutrients in food through the digestive tract.
- **Malar rash:** The medical term for the butterfly-shaped facial rash found in lupus.

Malignant: Cancerous.

- Mania: The high-energy phase of bipolar disorder.
- **Mast cells:** Specialized white blood cells that are found in connective tissue and contain histamine.
- Mastectomy: Surgical removal of the breast.
- **Meconium:** A dark greenish type of stool passed by a newborn during the first few days of life.
- **Medulloblastoma:** A type of malignant brain tumor that develops in the cerebellum. It is the most common type of brain tumor in children.
- **Melanin:** A brownish or dark reddish pigment that is the primary determinant of skin, hair, and eye color in humans.
- Melanocyte: A type of skin cell that produces melanin.
- **Melanoma:** The most serious form of skin cancer. Sunburn increases the risk of melanoma.
- **Melatonin:** A hormone produced in the pineal gland in the brain that regulates the sleep/wake cycle.
- **Meninges (singular, meninx):** The protective membranes that cover the brain and spinal cord.
- Meningioma: A type of brain tumor that starts in the meninges.
- **Meningitis:** Inflammation of the membranes that cover the brain and line the brain and spinal cord.
- **Metabolism:** The chemical changes in living cells in which new materials are taken in and energy is provided for vital processes,
- **Metastasis (plural, metastases):** The spread of a cancer from its original location to other organs or parts of the body.

- **Migraine:** A type of primary headache characterized by severe pain, nausea and vomiting, and sensitivity to light. It may occur on only one side of the head.
- **Milestone:** A physical development or accomplishment that most children reach within a specific age range.
- **Mixed state:** A condition in which a person with bipolar disorder has the energy of the manic phase of the disorder combined with the hopeless and sad mood of the depressed phase.
- **Mohs surgery:** A technique for removing skin cancers in very thin layers one at a time in order to minimize damage to healthy skin.
- **Monosomy:** A type of genetic disorder in which a cell contains only one copy of a particular chromosome instead of the normal two.
- **Mosaicism:** A condition in which a person has some body cells containing an abnormal number of chromosomes and other cells containing the normal number. Mosaicism results from random errors during the process of cell division that follows conception.
- **Motor neuron:** A type of cell in the central nervous system that controls the movement of muscles either directly or indirectly.
- **Mucous membrane:** Soft tissues that line the nose, throat, stomach, and intestines.
- **Multiple chemical sensitivity (MCS):** A controversial health condition related to a patient's belief that his or her symptoms are caused by exposure to environmental chemicals.
- Mutate: A change in the genetic material of an organism. Viruses can mutate rapidly.
- Mutation: A change in the genetic material of an organism.
- **Mycoplasma:** A very small bacterium that causes a mild but long-lasting form of pneumonia.
- **Myelin:** A fatty substance that insulates nerve fibers and allows for speedy and accurate transmission of nerve impulses.
- Myeloid: Relating to bone marrow.
- Myocardial infarction: The medical term for a heart attack.
- Myopia: The medical term for nearsightedness.
- Myosin: A protein involved in muscle movement.

**Myxedema:** A synonym for hypothyroidism. Myxedema coma is a condition in which a person with untreated hypothyroidism loses consciousness. It is potentially fatal.

## N

**Nebulizer:** A device that delivers medication in a fine spray or mist.

**Necrotizing:** Causing the death of soft tissue.

- **Neglect:** Failing to meet a child's basic needs for food, clothing, shelter, and medical care.
- **Negri bodies:** Round or oval bodies found within the nerve cells of animals infected by the rabies virus. They were first described by Dr. Adolchi Negri in 1903.
- Neonatal: The medical term for newborn.
- **Neural tube:** The medical term for the folds of tissue in the human embryo that eventually form the brain and spinal cord.
- **Neurologic:** Pertaining to the nervous system.
- **Neurologist:** A doctor who specializes in diagnosing and treating disorders of the nervous system.
- **Neurology:** The branch of medicine that studies and treats disorders of the nervous system.
- **Neurotransmitters:** Chemicals produced in the brain that transmit nerve impulses to other nerve cells and eventually to muscles.
- Nicotine: A chemical found in tobacco that acts as a stimulant in humans.

Nits: The eggs of lice.

- **Nodule:** The medical term for a small rounded lump of tissue.
- **Nondisjunction:** A genetic error in which one or more pairs of chromosomes fail to separate during the formation of germ cells, with the result that both chromosomes are carried to one daughter cell and none to the other.
- **Non-rapid eye movement (NREM) sleep:** The first phase of a sleep cycle, in which there is little or no eye movement.

- **Norepinephrine:** A brain chemical that affects a person's ability to pay attention.
- Nosocomial: Referring to a disease that a person gets while hospitalized.
- **Nucleotide excision repair (NER):** A mechanism that allows cells to remove damage caused by ultraviolet light to the cell's DNA.
- **Nucleotides:** The basic structural units of DNA and RNA, a cell's genetic material.

Nucleus: The innermost part of the lens of the eye.

Nymph: The second stage in the life cycle of the deer tick.

## 0

- **Obsession:** A recurrent, distressing, intrusive thought, image, or impulse.
- **Occult:** The medical term for a cancer that is too small to produce a visible tumor.
- **Oncogene:** A gene that has the potential to cause a normal cell to become cancerous.
- **Ophthalmia neonatorum:** The medical name for bacterial conjunctivitis in newborn babies caused by a sexually transmitted infection in the mother.
- **Ophthalmologist:** A doctor who specializes in diagnosing and treating eye disorders and can perform eye surgery.
- **Opportunistic infection:** An infection that occurs only in people with weakened immune systems.
- **Optician:** An eye care professional who fills prescriptions for eyeglasses and corrective lenses.
- **Optometrist:** An eye care professional who diagnoses refractive errors and other eye problems and prescribes corrective lenses.
- **Orphan drug:** A drug defined by the Food and Drug Administration (FDA) as intended to treat a disease or condition that affects less than 200,000 people in the United States, or a disease or condition that affects more than 200,000 people and there is no reasonable expectation that the company can recover the costs of developing the drug.

- **Orthokeratology:** A treatment for astigmatism that consists of wearing hard contact lenses overnight to reshape the cornea during sleep. The lenses are removed during the day.
- **Orthopaedics (also spelled orthopedics):** The branch of medicine that diagnoses and treats disorders of or injuries to the bones, muscles, and joints.
- **Ossicles:** A group of three small bones in the middle ear that transmit sound waves to the cochlea.
- **Osteopenia:** The medical name for low bone mass, a condition that often precedes osteoporosis.
- **Osteophyte:** A bony outgrowth or spur that develops in a joint affected by osteoarthritis. Osteophytes usually cause pain and limit the motion of the joint.
- **Osteoporosis:** A disease in which bones lose their density and are more likely to break or fracture under stress.
- Otitis: The medical term for inflammation of the ear.
- **Otolaryngologist:** A doctor who specializes in diagnosing and treating diseases of the ears, nose, and throat.
- **Otoscope:** An instrument with a light and magnifying lens that allows a doctor to examine the eardrum and ear canal.

## P

- **Paget disease:** A chronic disorder caused by a slow virus infection that results in deformed or enlarged bones.
- Pancolitis: Ulcerative colitis that affects the entire colon.
- **Pancreas:** A small organ that lies between the stomach and the liver and secretes insulin.
- **PANDAS disorders:** A group of disorders with psychiatric symptoms that develop in some children after strep throat or scarlet fever. The acronym stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections.
- **Pandemic:** A disease epidemic that spreads over a wide geographical area and affects a large proportion of the population.

- **Panic attack:** An episode of intense fear that lasts for several minutes and is accompanied by physical symptoms or temporary disturbances of thinking.
- Pap test: A screening test for cervical cancer devised by Giorgios Papanikolaou (1883–1962) in the 1940s.
- **Papule:** A small cone-shaped pimple or elevation of the skin.
- **Paradoxical undressing:** A symptom sometimes seen in people with moderate or severe hypothermia, thought to be caused by a malfunction of the hypothalamus. The person becomes confused, disoriented, and begins to remove clothing.
- **Paraplegia:** Paralysis that affects only the lower body.
- **Parotid glands:** Glands that produce saliva, located on each side of the face below and in front of the ear.
- **Pasteurization:** A process in which milk or fruit juice is partially sterilized by heating to a temperature that destroys disease bacteria without causing major changes in appearance and taste.
- **Pastia's lines:** Bright red lines that appear in the body folds of a patient with scarlet fever after the rash develops.
- Patent: The medical term for open or unobstructed.
- **Peak airflow meter:** A handheld device that asthma patients can use at home to monitor their lung capacity in order to treat the warning signs of an asthma attack as soon as possible.
- **Pelvic inflammatory disease (PID):** Inflammation of the uterus, fallopian tubes, and ovaries caused by chlamydia or gonorrhea. It can lead to permanent inability to have children if not treated.
- **Peptic ulcer:** The medical term for an ulcer in the digestive tract.
- **Perinatal:** Related to the period around the time of a baby's birth.
- **Periodontitis:** The medical term for gum disease that involves the connective tissue and bone beneath the gums.
- **Peritonitis:** Inflammation of the membrane that lines the abdominal cavity and covers some of the internal organs.
- **Pertussis:** The medical name for whooping cough.
- **Pervasive developmental disorder (PDD):** A diagnostic category for a group of childhood disorders characterized by problems in communication skills and social interactions.

- **Petechiae:** Tiny reddish or purplish spots in the skin caused by the breaking of small blood vessels during intense coughing or vomiting.
- **Pewter:** A metal made mostly of tin and small quantities of copper. Modern pewter is no longer made with lead.
- **Phacoemulsification:** A technique for removing cataracts by breaking up the lens of the eye with ultrasound waves and removing the pieces of the lens by suction.
- **Pharyngitis:** The medical term for sore throat.
- **Phenylalanine:** The amino acid that cannot be used by the bodies of people with phenylketonuria.
- Philadelphia chromosome: A genetic abnormality in chromosome 9 associated with CML. Its name comes from the location of the University of Pennsylvania School of Medicine, where it was discovered in 1960.
- **Phlegm:** Thick mucus secreted in the throat and lungs during an upper respiratory infection.
- **Phobia:** An unfounded or morbid dread of a specific object or situation that arouses feelings of panic.
- **Photophobia:** A feeling of discomfort or pain in the eyes during exposure to light.
- **Photopter:** A device positioned in front of a patient's eyes during an eye examination that allows the examiner to place various lenses in front of the eyes to determine the strength of corrective lenses required.
- **Phototherapy:** Method of treating skin disorders by exposing the affected skin to daylight or to specific wavelengths of visible or ultraviolet light.
- **Pica:** An abnormal craving for substances that are not normally considered food, like soil, chalk, paper, or ice cubes.
- Pinna: The visible part of the outer ear.
- Pitch: The highness or lowness of the voice or a musical note.
- **Pituitary gland:** A pea-sized gland located at the base of the brain behind the nose that secretes growth hormone and other hormones that affect sexual development and the body's response to stress.
- Plantar: Located on or referring to the sole of the foot.

- **Plaque (arterial):** A deposit of cholesterol and dead white cells along the inside wall of an artery.
- **Plaque (dental):** A film that forms on the surface of teeth containing bacteria, saliva, and dead cells.
- **Plasma:** The liquid part of blood, about 55 percent of blood by volume.
- **Platelet:** A small flat disk-shaped body in human blood that helps to form blood clots by sticking to other platelets and to damaged tissue at the site of an injury. Platelets are also called thrombocytes.
- **Plumbism:** The medical name for lead poisoning.
- **Polymyositis:** Inflammation of the muscles that causes weakness and difficulty in moving or swallowing.
- **Polyp:** A growth of tissue protruding from a mucous membrane such as the colon.
- **Polysomnograph:** A machine used in a sleep laboratory to monitor chest movement, air flow, brain waves, heart rhythm, and other data relevant to diagnosing sleep disorders.
- **Popcorn cell:** An abnormal cell found in nodular lymphocyte predominant Hodgkin disease (NLPHD).
- **Post-concussion syndrome (PCS):** A condition characterized by several weeks or months of headache following a head injury.
- **Postexposure prophylaxis (PEP):** A treatment given after exposure to the rabies virus. It consists of one dose of rabies immune globulin and five doses of rabies vaccine.
- **Postmortem:** Referring to the period following death.
- **Postpartum:** Referring to the period of time after giving birth.
- **Postpartum depression:** A type of depression that some women experience after the birth of a baby.
- **Postpartum psychosis:** A severe mental disorder in which the mother suffers from delusions or hallucinations.
- **Prader-Willi syndrome:** A rare genetic disorder characterized by mental retardation and an uncontrollable appetite for food.
- **Premutation:** An abnormally large number of repeated triplets in certain genes that does not cause obvious symptoms of a genetic disorder but can expand into a full mutation when transmitted to offspring.

Prenatal: Before birth.

- **Presbyopia:** Age-related farsightedness caused by loss of flexibility in the lens of the eye.
- **Pressure points:** Specific locations on the body where people with fibromyalgia feel pain even with light pressure.
- **Primary disease:** A disease that develops by itself and is not caused by a previous disease or injury.
- Prion: An abnormal infectious protein particle.
- **Proctitis:** The medical term for ulcerative colitis limited to the rectum.
- **Prodrome:** A period before the acute phase of a disease when the patient has some characteristic warning symptoms.
- **Progeria:** A disease characterized by abnormally rapid aging. The term can be used to refer specifically to Hutchinson-Gilford syndrome or to a group of diseases characterized by accelerated aging.
- **Progressive:** Referring to a disease or disorder that gets worse over time.
- **Prophylaxis:** The use of a medication or other therapy to maintain health and prevent disease.
- **Prostate:** A walnut-sized gland in males that secretes seminal fluid.
- **Protozoan (plural, protozoa):** A one-celled animal-like organism with a central nucleus enclosed by a membrane. Many protozoa are parasites that can cause disease in humans.
- **Pseudostrabismus:** A condition in which a child may seem to have strabismus because of certain facial features that change as the child's face matures.
- **Psychosis:** Severe mental illness marked by hallucinations and loss of contact with the real world.
- **Psychostimulant:** A type of drug that increases the activity of the parts of the brain that produce dopamine.
- **Pulp:** The soft living material in the center of a tooth that contains blood vessels and nerve endings.
- **Pupil:** The circular opening in the center of the iris.
- **Pus:** A whitish-yellow material produced by the body in response to a bacterial infection. It consists of tissue fluid and dead white blood cells.

**Pyelonephritis:** The medical term for a urinary tract infection that has spread from the bladder or other parts of the urinary tract upward to the kidneys.

## Q

- **Quadriplegia:** Paralysis that affects both arms and both legs. It is also known as tetraplegia.
- **Quarantine:** The practice of isolating people with a contagious disease for a period of time to prevent the spread of the disease.
- **Quinoa:** A plant grown in Peru and Bolivia for its edible seeds. It is high in protein and easy to digest.

### R

- **Radiologist:** A doctor who specializes in medical imaging techniques to diagnose or treat disease.
- **Radon:** A colorless and odorless gas produced by the breakdown of uranium known to cause lung cancer.
- **Rapid cycling:** Four or more episodes of illness within a 12-month period.
- **Rapid eye movement (REM) sleep:** The phase of a sleep cycle in which dreaming occurs; it is characterized by rapid eye movements.
- **Raynaud's phenomenon:** Discoloration of the fingers and toes caused by blood vessels going into spasm and decreasing the flow of blood to the affected digits.
- **Reactive hypoglycemia:** A condition in which a person develops hypoglycemia between two and five hours after eating foods containing high levels of glucose.
- **Rebound tenderness:** Pain experienced when the doctor releases pressure on the abdomen.
- **Rectum:** The lowermost portion of the large intestine, about 6 inches (15.2 centimeters) long in adults.

- **Reed-Sternberg cell:** An abnormal type of B lymphocyte that is found in classic Hodgkin lymphoma.
- **Reefer:** Another name for a marijuana cigarette.
- **Reflux:** The medical term for the backward flow of stomach acid from the stomach into the esophagus.
- **Refractive error:** A general term for vision problems caused by the eye's inability to focus light correctly.
- **Regurgitation:** Throwing up; effortless flow of undigested stomach contents back up the esophagus into the mouth.
- **Reiter's syndrome:** A type of arthritis than can develop in untreated people with chlamydia. It is characterized by inflammation of the genitals and the eyelids as well as sore and aching joints.
- Relapse: Recurrence of an illness after a period of improvement.
- **Remission:** A period in the course of a disease when symptoms disappear for a time.
- **Reservoir:** The term used by biologists for the natural host species of a disease organism. Bats are thought to be the reservoir of viral hemorrhagic fevers.
- **Resilience:** The capacity to recover from trauma and other stressful situations without lasting damage.
- **Resorption:** The removal of old bone from the body.
- **Retina:** The light-sensitive layer of tissue at the back of the eyeball.
- **Retinal detachment:** A disorder in which the retina pulls away from its underlying tissues at the back of the eye.
- **Rheumatoid factor (RF):** An antibody that attacks the body's own tissues that is found in some patients with rheumatoid arthritis and is measured as part of the diagnostic process.
- **Rheumatologist:** A doctor who diagnoses and treats diseases of the muscles, joints, and connective tissue.
- **Rheumatologist:** A doctor who specializes in diagnosing and treating arthritis and other diseases of the muscles and joints.
- **Rheumatology:** The branch of medicine that deals with disorders of the muscles, joints, and connective tissue.

- **Rhinitis:** The medical term for inflammation of the mucous tissues lining the nose. It can be caused by infections or chemical irritants as well as by allergies.
- **RNA virus:** A virus whose genetic material is composed of ribonucleic acid (RNA) and does not need DNA to copy itself and multiply.
- **Rocker-bottom feet:** Abnormally long and slender feet with pointed heels turned outward like the bottom rails of a rocker.
- **Rotator cuff:** A group of four muscles that attach the arm to the shoulder blade.

## S

Scald: A burn caused by steam or a hot liquid.

- Scaling: Scraping tartar away from the teeth around the gum line.
- **Scintigraphy:** A technique for detecting the location and extent of softtissue injury by injecting a small quantity of a radioactive element and following its distribution in the tissue with a scanner.
- **Sclera:** The opaque white portion of the eyeball.
- **Scleroderma:** A disorder of connective tissue characterized by thickening and tightening of the skin as well as damage to internal organs.
- **Sclerosis:** Hardening or scarring of tissue.
- Scoliosis: Abnormal curvature of the spine from side to side.
- **Scrapie:** A prion disease of sheep and goats.
- Sebum: An oily lubricant secreted by glands in the skin.
- **Secondary disease:** A disease that is caused by another disease or condition or by an injury.
- Selective serotonin reuptake inhibitors (SSRIs): A group of antidepressants that work by increasing the amount of serotonin available to nerve cells in the brain.
- Senile cataract: Another term for cataracts caused by the aging process.
- **Sepsis:** The presence of bacteria or their toxic products in the blood-stream or other tissues, leading to inflammation of the entire body.
- **Septum (plural, septa):** A partition that separates two cavities or chambers in the body.

- **Serotonin:** A brain chemical that influences mood, anger, anxiety, body temperature, and appetite.
- **Shingles:** A skin inflammation caused by reactivation of the chickenpox virus remaining in the nervous system.
- **Shock:** A medical emergency in which there is a drop in blood pressure and a reduced volume of blood circulating in the body.
- **Shunt:** A flexible plastic tube inserted by a surgeon to drain cerebrospinal fluid from the brain and redirect it to another part of the body.
- Sickle cell crisis: Sudden onset of pain and organ damage in the chest, bones, abdomen, or joints caused by defective red blood cells blocking blood vessels.
- **Sleep cycle:** A period of NREM sleep followed by a shorter phase of REM sleep. Most adults have four to six sleep cycles per night.
- **Slit lamp:** An instrument that focuses light into a thin slit. It is used by eye doctors to examine eyes for a wide variety of disorders.
- **Smoking cessation:** A term that refers to a product or program to help people quit smoking.
- **Snellen chart:** A series of letters arranged in lines on a chart to be viewed from a distance of 20 feet (6.1 meters) used to measure visual acuity (clearness of vision).
- **Soft palate:** The soft tissue at the back of the roof of the mouth that does not contain bone.
- **Solar keratoses (singular, keratosis):** Rough scaly patches that appear on sun-damaged skin. They are considered precancerous.
- **Spasticity:** Stiffness or spasms in the muscles.
- **Spectrum disorder:** A disorder whose symptoms vary in severity from one patient to the next.
- **Sphincter:** A ring-shaped muscle that can contract to close off a body opening.
- **Sphygmomanometer:** The device used to measure blood pressure. It consists of an inflatable cuff that compresses an artery in the arm. The doctor listens through a stethoscope as the air pressure in the cuff is released in order to measure the blood pressure.
- **Spirochete:** A spiral-shaped bacterium. Lyme disease is caused by a spirochete.

- **Spirometer:** A device that is used to test the air capacity of a person's lungs and the amount of air that enters and leaves the lungs during breathing.
- **Spleen:** An organ located behind the stomach that cleans old blood cells out of the blood and holds a reserve of red blood cells.
- **Sporadic:** Occurring at random.
- **Spore:** The dormant stage of a bacterium.
- **Sputum:** Mucus and other matter that is coughed or brought up from the lungs or throat.
- **Stacking:** Using several different types of steroids at the same time.
- Status epilepticus: An ongoing seizure that lasts longer than five minutes; it is a medical emergency.
- **Stem cell:** A type of unspecialized cell that can reproduce itself and differentiate into different types of specialized cells. Stem cells act as a repair system for the body.
- **Stenosis:** The medical term for abnormal narrowing of the opening of a blood vessel.
- **Stillbirth:** The birth of a baby that has died before or during delivery.
- **Stimulant:** Any drug or chemical that temporarily increases the user's awareness or alertness.
- **Stoma:** An opening made in the abdomen following surgery for digestive disorders, including colon cancer, that allows wastes to pass from the body.
- **Stomatitis:** The medical term for an inflammation of the mouth.
- **Strabismus:** A condition in which the eyes are not properly aligned with each other.
- Strain: A genetic variant or subtype of a bacterium.
- **Strawberry tongue:** A swollen and intensely red tongue that is one of the classic signs of scarlet fever.
- **Stress management:** Any set of techniques intended to help people deal more effectively with stress in their lives by analyzing specific stressors and taking positive actions to minimize their effects.
- **Stressor:** Any event or stimulus that provokes a stress response in a human or animal.

- **Stricture:** The medical term for an abnormal narrowing of a hollow organ like the bowel.
- **Substance P:** A chemical in the central nervous system that transmits pain signals back and forth between the brain and the rest of the body.
- **Sulcus (plural, sulci):** The space or crevice between a tooth and the surrounding gum tissue.
- Sun poisoning: A term sometimes used to refer to a severe reaction to sunburn, consisting of fever, chills, fluid loss, dizziness, and nausea.
- Sunsetting: A term used to describe a downward focusing of the eyes.
- **Surfactant:** A protein-containing substance secreted by cells in the lungs that helps to keep them properly inflated during breathing.

Surveillance: Monitoring of infectious diseases by public health doctors.

- Synapse: The medical term for specialized connections between nerve cells.
- **Syndrome:** A group of signs or symptoms that occur together and characterize or define a particular disease or disorder.
- **Synovium:** A type of tissue lining the joints that ordinarily secretes a fluid that lubricates the joints.

**Systemic:** Referring to a disease or disorder that affects the entire body. **Systolic blood pressure:** The blood pressure at the peak of each heartbeat.

## T

**Targeted therapy:** A newer type of cancer treatment that uses drugs to target the ways cancer cells divide and reproduce or the ways tumors form their blood supply.

Tartar: Hardened plaque.

- **Tendon:** A thick band or cord of dense white connective tissue that attaches a muscle to a bone.
- **Teratogen:** Any substance that causes birth defects in children. Alcohol is a teratogen.

**Testosterone:** The principal male sex hormone.

Thalassemia trait: A condition in which a person is missing one or two genes required to make the proteins in the alpha chains of the

hemoglobin molecule. The person does not have the symptoms of thalassemia but can pass the genetic deficiency to their children.

- **THC:** The abbreviation for delta-9-tetrahydrocannabinol, the main mind-altering chemical in marijuana.
- **Thrombus:** A blood clot that forms inside an intact blood vessel and remains there.
- **Thymus:** A small organ located behind the breastbone that is part of the lymphatic system and produces T cells.
- **Thyroid storm:** A medical emergency marked by a rise in body temperature as well as other symptoms caused by untreated hyperthyroidism.
- **Thyroiditis:** Inflammation of the thyroid gland.
- **Tic:** A sudden repetitive movement or utterance. Tourette syndrome is considered a tic disorder.
- **Tick:** A small bloodsucking parasitic insect that carries Lyme disease and several other diseases.
- Tinnitus: The medical term for ringing in the ears.
- **Tolerance:** The need for greater and greater amounts of a drug to get the desired effects.
- **Tonometer:** An instrument used by an ophthalmologist to measure the pressure of the fluid inside the eye.
- **Tonsillectomy:** Surgical removal of the tonsils.
- **Topical:** Referring to a type of medication applied directly to the skin or outside of the body.
- **Tourette syndrome:** A neurological disorder characterized by recurrent involuntary body movements and repeated words or grunts.
- **Toxin:** A poisonous substance produced by a living cell or organism.
- **Trachoma:** An infectious disease of the eye caused by chlamydia bacteria that can lead to blindness if untreated.
- **Traction:** The use of braces, casts, or other devices to straighten broken bones and keep them aligned during the healing process.
- **Transdermal:** Referring to a type of drug that enters the body by being absorbed through the skin.

- **Transient ischemic attack (TIA):** A brief stroke lasting from a few minutes to twenty-four hours. TIAs are sometimes called ministrokes.
- **Translocation:** A genetic error in which a part of one chromosome becomes attached to another chromosome during cell division.
- Trauma: A severe injury or shock to a person's body or mind.
- **Tremor:** Trembling or shaking caused by a physical disease.
- **Triglyceride:** A type of fat made in the body.
- **Triplet:** In genetics, a unit of three nucleotides that starts or stops the production of a specific protein. Triplets are also called codons.
- **Trismus:** The medical name for the spasms of the jaw muscles caused by tetanus.
- **Trisomy:** A type of genetic disorder in which a cell contains three copies of a particular chromosome instead of the normal two.
- **Triticale:** A grain that is a cross between wheat and rye, first grown in Scotland and Sweden in the nineteenth century.
- **T-score:** The score on a bone density test, calculated by comparing the patient's bone mineral density to that of a healthy thirty-year-old of the same sex and race.
- **Tumor:** An abnormal mass or growth of tissue that may be either cancerous or noncancerous.
- **Tumor markers:** Substances found in blood, urine, or body tissues that can be used to detect cancer.

## U

- **Ureter:** A muscular tube that carries urine from the kidney to the bladder.
- **Urethra:** The tube that allows urine to pass from the bladder to the outside of the body.
- **Urologist:** A doctor who specializes in diagnosing and treating disorders of the kidneys and urinary tract.
- Urticaria: The medical term for hives.
- **Uveitis:** Inflammation of the interior of the eye.

**Uvula:** A triangular piece of soft tissue located at the back of the soft palate.

V

- **Vasoconstriction:** A narrowing of the blood vessels in response to cold or certain medications.
- **Vector:** An insect or other animal that carries a disease from one host to another.
- **Ventricle (brain):** One of four hollow spaces or cavities in the brain that hold cerebrospinal fluid.
- Ventricle (heart): One of the two lower chambers of the heart.
- Verruca (plural, verrucae): The medical term for a wart.
- **Vertebra (plural, vertebrae):** One of the segments of bone that make up the spinal column.
- **Vesicle:** A small blister or sac containing fluid.
- **Vestibular system:** The group of organs in the inner ear that provide sensory input related to movement, orientation in space, and balance.
- **Villi (singular, villus):** Small finger-like projections along the walls of the small intestine that increase the surface area of the intestinal wall.
- **Viral load:** A measure of the severity of HIV infection, calculated by estimating the number of copies of the virus in a milliliter of blood.
- Virilization: The development of male sexual characteristics in females.
- **Vital signs:** Measurements taken to evaluate basic body functions. They are temperature, pulse rate, blood pressure, and breathing rate.
- **Vocal folds:** Twin folds of mucous membrane stretched across the larynx. They are also known as vocal cords.

W

Wasting: Loss of lean muscle tissue.

Werner syndrome: A genetic disease characterized by accelerated aging.

Wheal: A suddenly formed flat-topped swelling of the skin; a welt.

- **Wheezing:** A continuous harsh whistling sound produced by the airways of an asthma patient when the air passages are partly blocked.
- Whipple triad: A group of three factors used to diagnose hypoglycemia: symptoms; blood sugar measuring below 45 mg/dL for a woman and 55 mg/dL for a man; and rapid recovery following a dose of sugar.
- **Window period:** The period of time between a person's getting infected with HIV and the point at which antibodies against the virus can be detected in a blood sample.
- **Withdrawal:** A collection of signs and symptoms that appear when a drug (including alcohol, caffeine and nicotine) that a person has used for a long time is suddenly discontinued.
- **With-the-rule astigmatism:** A type of astigmatism in which the eye sees vertical lines more clearly than horizontal lines.
- **Wood's lamp:** A special lamp that uses ultraviolet light to detect certain types of skin infections and infestations.



**Xerostomia:** The medical term for dry mouth.



Yaws: A tropical, bacterial infection of the skin, bones, and joints.

## Z

**Zoonosis (plural, zoonoses):** A disease that can be transmitted from animals to humans.

# A



**Also Known As** Dwarfism, short-limb dwarfism

**Cause** Genetic mutation

#### Symptoms

Short stature, short arms and legs, large head

Duration Lifelong



#### Definition

Achondroplasia, or short-limb dwarfism, is the most common form of abnormally short stature in adults. It is caused by a mutation in a single gene on chromosome 4 that regulates the conversion of cartilage to bone. This gene is the only gene that is known to be associated with achondroplasia.

#### Description

Achondroplasia is basically a disorder of bone development. The skeleton of a human fetus is composed primarily of cartilage, a dense and somewhat elastic form of connective tissue that gradually turns to bone during normal development. In a person with achondroplasia, a gene that is involved in the process of bone formation produces too much of a protein that limits bone growth. As a result, the person with achondroplasia has unusually short bones in the arms and legs and other skeletal abnormalities. They also usually have difficulties with posture, joint disorders, and breathing problems in later life.

#### Demographics

Researchers estimate that achondroplasia occurs in one in every 15,000 to 40,000 live births. About 20 percent of cases are children who have one parent with achondroplasia; however, 75–80 percent of cases involve new



X ray showing the skull and spine of a person with achondroplasia. SCOTT CAMAZINE / PHOTO RESEARCHERS, INC.

mutations of the gene responsible for the disorder. These new mutations are more likely to occur in the sperm of fathers over 35; the mother's age does not matter, as far as is presently known.

The disorder affects both sexes and all races equally.

The average adult height of people with achondroplasia is 4 feet 4 inches (1.3 meters) for men and 4 feet one-half inch (1.24 m) for women. The shortest living person with achondroplasia as of 2008 was Jyoti Amge, a teenager from Nagpur, India, who stands 23 inches (58 centimeters) tall and weighs 11 pounds (5 kilograms).

#### **Causes and Symptoms**

Achondroplasia is caused by a mutation in the FGFR3 gene on chromosome 4. A normal gene helps the body convert cartilage to bone. The mutation in the gene related to achondroplasia results in severely shortened bones and weak muscle tone in the body.

The symptoms of achondroplasia are usually obvious by the first year of life. They include:

- Poor muscle tone.
- Slowness in learning to walk; a child with achondroplasia may not walk until some point between 24 and 35 months.
- Abnormal skull structure leading to frequent ear infections, apnea (temporary slowing or stopping of breathing), and overcrowding of the teeth.
- Distinctive facial features, particularly a prominent forehead and underdevelopment of the nose and midface.
- Greater than normal separation of the little finger from the ring finger, giving the hand a distinctive trident (three-pronged) shape.
- A tendency toward obesity.
- Postural problems, including a pronounced curvature of the spine in the lower back and bowed legs.
- Back and leg pain in adult life.

#### **Billy Barty**

Billy Barty (1924–2000) was an Italian-American film actor who played in a number of movies from the 1930s through 2001. He also had roles in several television series, from *Alfred Hitchcock Presents* and *Peter Gunn* to *Little House on the Prairie* and *Frasier*. Born William John Bertanzetti in Millsboro, Pennsylvania, Barty became a noted spokesperson and activist for persons with dwarfism.

In 1957 Barty gathered a group of others diagnosed with achondroplasia to meet with him in Reno, Nevada, to start an organization that would offer support and information to people with dwarfism and their families. Twenty-one people came to the meeting and formed Little People of America (LPA), an organization that has over 6,000 members as of the early 2000s.

Barty was once asked in the 1970s why he worked so tirelessly to end stereotypes and ridicule of people with dwarfism. He said, "Most of us with dwarfism prefer to be described as 'Little People.' And please, put the emphasis on the word *People*. We did not spring from the pages of a story book or emerge from an enchanted forest. We are not magical beings and we are not monsters. We are parents and sons and



*Billy Barty on Hollywood Boulevard's Walk of Fame.* AP IMAGES.

daughters. We are doctors and lawyers and realtors and teachers. We dream, cry, laugh, shout, fall in love, and make mistakes. We are no different from you."

#### Diagnosis

Babies with achondroplasia can be diagnosed before birth by ultrasound measurements of the growth of their long bones and head size. After birth, the diagnosis is usually based on x-ray studies of the child's bones and head size. The x-ray images usually reveal a small skull base, shortened growth plates in the long bones, square-shaped long bones, and normal-sized bones in the trunk area.

A genetic test can also be performed to confirm the diagnosis in children whose symptoms may be less clear.

#### Treatment

The management of achondroplasia includes careful monitoring of the child's growth, head size, and weight pattern. There are special growth charts that doctors use to evaluate the rate of growth in children with achondroplasia. It is particularly important to prevent obesity if possible. In addition, children with achondroplasia need social support because of ongoing prejudice against people with dwarfism. Organizations such as Little People of America can offer helpful advice on the education and other future plans of children with achondroplasia.

Surgery may be performed if necessary to relieve pressure on the spinal cord or the brain. In some cases, there may be a buildup of fluid in the brain (hydrocephalus) that needs to be drained surgically. The child's tonsils and adenoids may be removed in order to lower the risk of apnea. The shape of bowed legs can be corrected surgically, but doctors disagree about the value of surgery intended to lengthen the legs.

Children with achondroplasia may need extra dental work because of the mismatch between the size of their teeth and the size of the jaw. In addition, they should be watched carefully for recurrent ear infections in order to minimize the risk of deafness and later learning difficulties.

There is disagreement among doctors as of the early 2000s regarding treatment with human growth hormone. It is considered an experimental treatment. Many doctors are concerned that using growth hormone in children with achondroplasia will lead to abnormal bone deposits and worsening of the spinal curvature.

#### Prognosis

The prognosis of a baby with achondroplasia depends on whether the defective gene is inherited from only one parent or from both. A child who inherits the gene from both parents will die before birth or shortly after birth from respiratory failure. About 3 percent of children with achondroplasia die suddenly and unexpectedly during the first year of life, usually from compression of the spinal cord.

A child who has one normal copy of the gene will usually have normal intelligence and a normal life expectancy in spite of bone and joint problems.

#### Prevention

There is no known way to prevent achondroplasia because the gene responsible for the condition can undergo mutation in families with no

#### WORDS TO KNOW

**Apnea:** A condition in which breathing slows or stops for short periods of time.

- **Cartilage:** A dense elastic tissue that forms most of the skeleton in the human fetus but is gradually replaced by bone during normal development.
- **Growth plate:** A cartilage plate in the long bones of children where the lengthening of bone takes place.
- **Hydrocephalus:** An abnormal increase of cerebrospinal fluid within the brain that may cause enlargement of the skull.
- **Mutation:** A change in the genetic material of an organism.

history of the disorder, and 75–80 percent of cases result from new mutations.

#### The Future

Although new mutations of the gene associated with achondroplasia cannot always be foreseen, people who have the condition should seek genetic counseling before marriage. If the partner does not have the mutation, there is a 50 percent chance with each pregnancy that the child will have achondroplasia. If both spouses have the mutation, they have a 25 percent chance of a normal child with each pregnancy, a 50 percent chance that the child will have achondroplasia, and a 25 percent chance that the child will have achondroplasia, and a 25 percent chance that the child will die before or shortly after birth.

SEE ALSO Childhood obesity; Ear infection; Gigantism; Hydrocephalus

#### For more information

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## Acid Reflux

See Gastroesophageal reflux disease.



## Definition

Acne is a skin disorder resulting from the blockage of hair follicles in the skin. Excess cells produced in the follicle combine with sebum, an oily substance secreted by glands connected to the follicle. When a plug forms in the follicle and is infected with bacteria, a pimple erupts.

#### Description

Acne is a skin problem commonly associated with teenagers, but it can also occur in children, adults in their thirties or forties, and women around the time of menopause. It is most likely to appear on the parts of the body with the largest number of hair follicles—the face, chest, upper back, and (in some people) upper arms. Some forms of acne are

#### Also Known As

Acne vulgaris, zits, pimples, blemishes

#### Cause

Interaction of hormones, oil glands, and hair follicles

#### Symptoms

Whiteheads, blackheads, inflamed eruptions, scarring

**Duration** Months to years

#### Acne

relatively mild. The less severe form consists of comedones, which are hair follicles blocked by plugs of sebum. If the comedo is open to the air on the skin surface, the sebum grows darker, giving the comedo a blackish appearance. This type of comedo is called a blackhead; its dark color is not caused by dirt. If the comedo is not open, the sebum inside produces a whitish bump called a whitehead. Both whiteheads and blackheads can remain in the skin for



weeks. Bacteria that normally live on the skin can grow inside the plugged follicles. The bacteria then secrete various chemicals that prompt an inflammatory response from the person's immune system. The plugged follicle may eventually burst, allowing the bacteria, the sebum, dead skin cells, and dead white blood cells to leak into nearby skin, causing reddened papules (small pink bumps), pus-filled pimples, or cysts (deep pus-filled lesions that are sore to the touch and can cause scarring).

#### *Illustration of a normal bair follicle and an inflamed one results in acne.* © NUCLEUS MEDICAL ART, INC. / ALAMY.

#### Demographics

According to the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), about 80 percent of people in the United States develop acne at some point between the ages of eleven and thirty. Boys are more likely than girls to develop acne during adolescence; however, among adults, women are more likely than men to suffer from acne. Twelve percent of women and 5 percent of American men at age twenty-five have acne; at age forty-five, 5 percent of both men and women still have acne. Some older women are troubled by acne again during menopause. Acne is equally common among people of all races and ethnic groups in the United States.

#### **Causes and Symptoms**

Acne is thought to result from the interaction of four factors that are most common in adolescents but can occur in adults as well. The first is the rise in production of hormones known as androgens, which are hormones produced by the adrenal glands (located on top of the kidneys) and are present in women as well as in men. The hormonal



Acne on a person's chin. © NAS MEDICAL / ALAMY. increase triggers the rapid multiplication of the cells lining the hair follicles on the face, chest, upper back, and shoulders. When these cells are produced faster than the follicle can shed them, they form a plug that blocks the follicle and keeps the sebum produced by the glands connected to the follicle from reaching the surface of the skin. The mixture of oily sebum and dead skin cells in the plug allows bacteria known as *Propionibacterium acnes*, which normally live on the skin, to grow inside the follicle. The che-

micals produced by the bacteria then trigger an inflammatory response from the body's immune system. White blood cells are drawn to the area around the plugged follicle to fight the bacteria. If the plugged follicle bursts, the inflammation may spread, leading to the pus-filled pimples and cysts of severe acne. Some people are more likely than others to develop acne.

Risk factors include:

- Heredity. People with a family history of acne are more likely to develop acne at a relatively early age and to have more severe breakouts.
- Changes in the body's hormonal levels. Pregnant women, women in the week preceding the menstrual period, women in early menopause, and teenagers of either sex are more likely to develop acne. People who use steroids for bodybuilding, medications containing cortisone, antiepileptic medications, or lithium are also more likely to have acne.
- Exposure to greasy or oily substances (deep-fat fryers or similar cooking equipment, lubricating oils and grease guns, oil-based paints) at home or in the workplace.
- Clothing or athletic equipment (backpacks, shoulder straps, helmets, headsets, etc.) that puts pressure on skin or rubs against it.
- Climate. People who live in locations with high humidity or high levels of air pollution are more likely to develop acne.

Mild acne is not painful; the pustules and cysts of severe acne, however, may be sore to the touch. In addition, the psychological effects of acne can be very painful for many patients, particularly adolescents.

#### Diagnosis

In most cases the doctor diagnoses acne on the basis of the appearance of the skin. Primary care doctors can usually treat milder cases of acne. People with more severe cases are usually referred to a dermatologist, who is a doctor who specializes in diagnosing and treating skin disorders. In a very few cases, women whose skin problems may be related to abnormally high levels of androgens may be given a hormone test.

#### Treatment

Treatments for acne work in one or more of four ways:

- lowering the skin's production of sebum
- speeding up the removal of dead skin cells
- fighting bacterial infection
- reducing the skin's inflammatory response to infection

Some medications used to treat acne are topical (applied to the skin) while others are taken by mouth. Some can be purchased over the counter, but others require a prescription from the doctor.

The specific medications or other treatments that a doctor might recommend depend on the severity of the acne, the extent of scarring, and the possibility of side effects for specific patients. Mild acne is commonly treated with topical medications in soap, cream, or lotion form, most of which do not require a prescription. They include various combinations of sulfur, benzoyl

#### Self-Care for Acne

To care for skin troubled by acne, people should:

- Cleanse the skin gently twice a day with a cleanser recommended by the doctor, and rinse the cleanser thoroughly.
- Wash the face after exercise or other activities that lead to heavy sweating.
- Wash the hair regularly, particularly if the facial skin is oily. Avoid rubbing or touching the face, or squeezing the pimples. Pinching or picking at acne blemishes can lead to scarring.
- Men who need to shave should shave carefully and use the type of razor that works best for them, whether electric or safety blade models. They should be careful when shaving to avoid nicking or cutting the pimples.
- Avoid unnecessary sun exposure, as many acne medications increase the risk of sunburn.
- Women who use cosmetics should choose them carefully and avoid oilbased products. All makeup should be removed before bedtime.
- Wear loose-fitting clothing that does not trap sweat or rub against the skin helps to reduce the risk of acne breakouts on the neck, upper arms, and upper back.

peroxide, salicylic acid, and a few other drugs to dry up excess sebum, kill bacteria, and speed up removal of dead skin cells. These nonprescription products may take about eight weeks to produce results. Moderately severe acne may be treated with prescription medications, including antibiotics to be taken by mouth along with using topical creams, lotions, or gels that also require a prescription. These products may cause stinging, reddening, or peeling of the skin in some people; however, patients usually start seeing improvement in about four weeks.

Patients with severe cystic acne are usually referred to a dermatologist for specialized treatment. A drug that is often prescribed for severe acne is Accutane, a drug derived from vitamin A that shrinks the size of the oil glands that produce sebum. Accutane must be taken only under careful supervision by a doctor, however, as it can cause serious side effects, including depression and an increased risk of a disease called irritable bowel syndrome. In addition, Accutane cannot be given to women who may become pregnant because it can cause birth defects in children. Other treatments for acne include phototherapy, which is the use of light waves to kill bacteria; and laser therapy, which helps to dry up the oil glands in the skin.

People with severe scarring from acne can have their scars treated with dermabrasion or laser therapy to make them less noticeable. Very large scars can be removed surgically if necessary.

#### Prognosis

The prognosis for acne depends on its severity. Most people's skin clears up by the early adult years, and newer techniques for treating scars lead to good results for most people. Nonetheless, some people have longstanding psychological problems—usually social isolation—as the result of severe acne in adolescence.

#### Prevention

Acne is difficult to prevent entirely, particularly for people who have inherited a family tendency to develop severe acne in adolescence. However, careful attention to proper skin care can reduce the severity and frequency of breakouts.

#### The Future

Current research focuses on possible ways to prevent skin inflammation caused by *Propionibacterium acnes*. Scientists in Germany have recently identified the bacterium's genome (genetic information), which may lead to the development of new medications to eliminate the bacterium from acne-troubled skin. Other research is directed toward developing lasers that can treat acne scarring with less damage to the outer layers of skin.

#### WORDS TO KNOW

- **Androgens:** The group of male sex hormones secreted by the outer portion of the adrenal gland; they are found in women as well as men.
- **Comedo (plural, comedones):** The medical term for a whitehead or blackhead.
- **Dermabrasion:** Technique for making acne scars less noticeable by removing the top layer of skin with a rapidly rotating wire brush or a sandy material.
- **Dermatologist:** Doctor who specializes in treating disorders of the skin.

- **Follicle:** Small canal in the skin surrounding the root of a hair.
- **Phototherapy:** Method of treating skin disorders by exposing the affected skin to daylight or to specific wavelengths of visible or ultraviolet light.
- **Sebum:** An oily lubricant secreted by glands in the skin.
- **Systemic:** Referring to a disease or disorder that affects the entire body. Acne is not a systemic disease.
- **Topical:** Referring to a medication applied to the skin or the outside of the body.

SEE ALSO Irritable bowel syndrome; Sunburn

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## AIDS

## Definition

AIDS is the end stage of an infectious disease caused by the human immunodeficiency virus, or HIV. The virus damages the person's immune system, leaving them vulnerable to certain cancerous tumors and increasingly severe opportunistic infections. HIV can be transmitted whenever a body fluid containing the virus (especially blood or sexual fluids) comes into contact with a mucous membrane (soft tissues lining various body cavities, like the mouth or vagina) or the bloodstream itself. A person can get AIDS through sexual intercourse, anal or oral sex, childbirth, breastfeeding, blood transfusion, or sharing hypodermic needles.

### Description

AIDS is now considered a pandemic because it has spread to every country in the world. According to the World Health Organization (WHO), 33.2 million people around the world were living with HIV infection in 2007; 2.1 million people died in that year from the disease, 330,000 of them children. Scientists think that the virus that causes AIDS originated somewhere in the African rainforest as an infection of chimpanzees and Old World monkeys. At some point in the twentieth century it jumped the species barrier from monkeys into humans. AIDS was first defined as an epidemic human disease in June 1981 by the Centers for Disease Control and Prevention (CDC). The virus that causes AIDS was identified by two teams of French and American scientists in 1983–1984.

HIV infection progresses in stages as the virus gradually weakens the body's immune system. It takes an average of eleven years for HIV infection to progress to AIDS. AIDS is diagnosed when the count of certain white blood cells in the patient's blood drops to a critical level or the patient develops life-threatening tumors or opportunistic infections.

#### Also Known As

Acquired immunodeficiency syndrome, HIV infection

#### Cause

Human immunodeficiency virus (HIV)

#### Symptoms

Fever, headache, swollen glands (early); diarrhea, weight loss, night sweats, infections (later)

## Duration

Months to years



Foot showing sores from Kaposi's sarcoma, a cancer commonly found in AIDS patients. COURTESY OF CENTERS FOR DISEASE CONTROL AND PREVENTION (CDC).

In the early stage of HIV infection, the patient may have no symptoms at all or a mild flu-like illness with fever and headache within a few days or weeks of getting infected. These symptoms usually go away without treatment and the person feels normal, even though he or she can transmit the infection to others. The infected person may continue to feel well for a period ranging from a few months to several years.

#### Demographics

Worldwide, about 0.6 percent of the population is infected with HIV, or about 35 million people. Ninety-five percent of these cases are in Africa or southeastern Asia. About 25 million people have died of AIDS since 1981, making the disease one of the deadliest pandemics in history. In the United States, the CDC's recently revised estimates indicate that about 945,000 people have been diagnosed with AIDS since 1981, and about 1.2 million are currently living with HIV infection. About a quarter of these people are unaware that they are infected. The CDC estimates that there are about 56,300 new cases of HIV infection in the United States each year.

In terms of specific groups within the United States:

• Males account for 74 percent of persons with HIV infection. Worldwide, the figure is 50 percent.

## **Ryan White**

Ryan White (1971–1990) was a teenager with hemophilia who became infected with AIDS from contaminated blood in 1984. He was expelled from his school at the time of his diagnosis because HIV infection was not well understood at the time and was thought to be primarily a disease of male homosexuals. At the time that White was diagnosed as the result of an opportunistic lung infection, his doctors thought that he had only six months to live. His family moved to another town in Indiana so that he could finish high school in a community that accepted him. White died in April 1990 just before he was ready to graduate from high school. In the six years between his diagnosis and his death, he became a spokesperson for people with AIDS, appearing on news programs and testifying before a national commission on the disease.

Ryan White's legacy is the Ryan White CARE Act, which provides federal funds for the care of people living with HIV/AIDS. The act was passed in 1990 and reauthorized in 1996, 2000, and 2006. It presently provides care for about 500,000 people in



*Ryan White in 1985.* AP IMAGES. the United States at a cost of over \$2.1 billion per year.

- In terms of race or ethnicity, 47 percent of persons with HIV infection are African American, 34 percent are Caucasian, 17 percent are Hispanic, and 2 percent are Native American or Asian American.
- In terms of method of transmission, 50 percent of infected persons are men who had sex with men; 33 percent had high-risk heterosexual sex; 13 percent are injection drug users; and the remainder are people who engaged in more than one high-risk behavior.
- In terms of age group, 1 percent of infected persons are under thirteen years of age; 15 percent are between the ages of thirteen and twenty-four; 26 percent are between the ages of twenty-five and thirty-four; 32 percent are between the ages of thirty-five and fortyfour; 20 percent are between the ages of forty-five and fifty-four; 8 percent are fifty-five or older.

#### **Causes and Symptoms**

The cause of AIDS is infection with human immunodeficiency virus or HIV. HIV is a virus that reproduces by inserting its own genetic material into a type of white blood cell called a CD4 lymphocyte. When the virus copies break out of the infected white blood cell, they attack other CD4 cells and the cycle repeats. Eventually so many of the white blood cells have been destroyed that the body's immune system is weakened and the person can no longer fight off opportunistic infections. The person may also develop certain cancers associated with a weakened immune system.

In the early stage of HIV infection, the patient may have no symptoms or else a mild flu-like illness with headache, fever, and swollen lymph nodes. Some patients have a second set of symptoms, often years after the initial infection, characterized by diarrhea, weight loss, shortness of breath, and coughing. To meet the official definition of AIDS, however, the patient must develop an opportunistic infection and have a CD4 cell count below 200. Opportunistic infections include diseases like toxoplasmosis, yeast infections of the esophagus, pneumonia caused by an organism known as *Pneumocystis*, and various disorders of the digestive tract caused by parasites. Cancers associated with AIDS include Kaposi sarcoma, a skin cancer; and non-Hodgkin lymphoma.

Other symptoms of full-blown AIDS include:

- Soaking night sweats
- Fever over 100°F (37.7°C) that lasts for several weeks
- Headaches
- · Blurred vision or other vision problems
- Chronic diarrhea
- Swelling in the lymph nodes that lasts for three months or longer

#### Diagnosis

The diagnosis of HIV infection and AIDS is complicated by the fact that many people are afraid to be tested for the disease. They may fear that a positive test will lead to the loss of housing, jobs, relationships, or the chance to complete their education. Because many infected persons put off getting tested and telling their partners, the disease continues to spread. In 2006, the CDC recommended routine HIV screening for all adults, adolescents, and pregnant women within health care settings, not just those considered to be high-risk. Testing for HIV is a two-step process. The first test is a screening test, which usually involves taking a sample of the patient's blood. There are newer screening tests that can use a sample of the person's urine or saliva. These rapid screening tests look for antibodies to the HIV virus and give results in about twenty minutes. If the person tests positive for HIV infection, a more specific second test, called a Western blot test, is performed. This test uses a blood sample to identify antibodies against HIV.

In 1996 the Food and Drug Administration approved a test kit that people can use at home called the Home Access HIV-1 Test. The person pricks their finger on a special blotting card and mails it back to the company. The sample is identified only by a code number, which allows the person to remain completely anonymous. The test costs about \$45.00 and results are available in seven days.

An important point to keep in mind is that it may take the body several weeks to three months after a person is infected to produce enough antibodies to HIV to be detected by a blood test. This period of time is called the window period. A person who tests negative for HIV infection after high-risk behaviors should wait three months and have another blood test to make sure they are not infected.

#### Treatment

There currently is no cure for HIV infection or AIDS. When a person tests positive for HIV infection, the doctor will measure the amount of virus in the patient's blood. This level is called the viral load. The viral load helps the doctor to decide when to start drug treatment for HIV. The current method of treatment is called highly active antiretroviral therapy or HAART. Introduced in 1996, HAART consists of combinations of three or more different drugs from two or more of the classes of antiretroviral drugs presently available. HAART is not a cure for AIDS, but it reduces the viral load, improves the patient's overall quality of life, and extends life expectancy.

HAART has several drawbacks. First, it can be very expensive form of treatment. In addition, the drugs used in HAART have troublesome side effects; as a result, some AIDS patients simply stop taking their medications. Last, some patients develop resistance to the antiretroviral drugs and no longer respond to treatment especially if the patient does not follow the treatment course and misses dosages. The doctor can sometimes switch one of the drugs in the patient's combination to another drug within the same class or another class.

## Prognosis

AIDS cannot be cured. Without treatment, HIV infection progresses to AIDS in an average of eleven years. After diagnosis with AIDS, the patient has a life expectancy of just over nine months without treatment. A person diagnosed with HIV infection who begins treatment with HAART can expect to live about twenty years or more after the diagnosis. Unfortunately, about half of patients who begin treatment with HAART fail to benefit from it as much as they had hoped and discontinue it.

### Prevention

There is no vaccine against HIV infection; moreover, it is unlikely that an effective vaccine will be developed in the foreseeable future because the virus that causes AIDS mutates so rapidly. Although various vaccines against HIV have been tested by the National Institutes of Health since 1996, none have so far been approved for use outside clinical trials.

People can lower their risk of HIV infection by taking the following precautions recommended by the CDC:

- Limit sexual activity to a single partner who is known to be uninfected and is faithful.
- Use a condom when having sex with anyone whose HIV status is unknown.
- Do not share needles or inject illegal drugs.
- Do not exchange sex for drugs.
- Health care workers should follow guidelines for protecting against needle sticks and other accidental exposures to body fluids that may be contaminated with HIV.
- Get tested for HIV infection after engaging in high-risk activities; if the test results are positive, inform all current sexual partners.

#### The Future

The demographics of HIV infection within the United States are changing somewhat, with women accounting for more new cases than in 1998. A worrisome new trend is the return and increase of high-risk

## WORDS TO KNOW

- **Highly active antiretroviral therapy (HAART):** An individualized combination of three or more antiretroviral drugs used to treat patients with HIV infection.
- **Opportunistic infection:** An infection caused by an organism that does not cause disease in a person with a healthy immune system.
- **Pandemic:** An infectious disease that spreads across a large region or even worldwide.

- **Viral load:** A measure of the severity of HIV infection, calculated by estimating the number of copies of the virus in a milliliter of blood.
- Window period: The period of time between a person's getting infected with HIV and the point at which antibodies against the virus can be detected in a blood sample.

behaviors among men who have sex with men. This trend appears to have been triggered by the spread of methamphetamine addiction from the West Coast to the Eastern Seaboard over the past several years.

Researchers at the National Institutes of Health are continuing to test various vaccines against HIV. They are also working on developing new antiretroviral drugs for patients who have developed resistance to those presently in use.

**SEE ALSO** Depression; Gonorrhea; Hemophilia; Lymphoma; Pneumonia; Syphilis; Toxoplasmosis

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## Alcoholism

### Definition

Alcoholism is a chronic (long-term) disease in which people become physically dependent on alcohol and cannot control how much they drink even though their drinking is damaging their health, schoolwork or job performance, and friendships or family relationships. It is a progressive disease, which means that without treatment, it gets worse over time.

Some doctors use the term "alcohol abuse" to describe heavy drinking that causes problems in the person's life but has not yet led to physical dependence on alcohol or complete loss of control over drinking.

Binge drinking refers to consuming a number of drinks in a row on a single occasion, usually five drinks for men and four for women. A drink is defined as one 12-ounce beer, one 4- to 5-ounce glass of wine, or one mixed drink containing 1.5 ounces of whiskey or other liquors. Some binge drinkers may claim that they do not have a problem with alcohol because they do not drink every night.

#### Also Known As

Alcohol abuse, alcohol dependence, problem drinking

#### Cause

Combination of genetic, social, and personal psychological factors

#### Symptoms

Craving for alcohol, physical dependence on it, loss of control over drinking, tolerance

#### Duration Years

#### Alcoholism

Image of a human liver with fibrosis, the formation of tough scar tissue in the liver, which can lead to cirrhosis. © NUCLEUS MEDICAL ART, INC./ ALAMY.



## Description

Alcoholism is a serious disease not only for the damage it does to the health and lives of alcoholics but also to the burden it places on society as a whole. It is the third leading cause of preventable death in the United States, after smoking and obesity. Alcoholism is responsible for 85,000 deaths in the United States each year. About one in every twelve adults in the United States—17.6 million people—abuse alcohol or are dependent on it.

Around the world, alcoholism is responsible for a percentage of various disease conditions:

- Cirrhosis of the liver: 32 percent
- Motor vehicle crashes: 20 percent
- Cancer of the esophagus: 29 percent
- Liver cancer: 25 percent
- Stroke: 10 percent
- Homicide: 24 percent
- Suicide: 11 percent

Women who drink during pregnancy are at risk of having children who are mentally retarded and underweight at birth. This condition,

## **Two Screeners for Alcoholism**

Two brief screening tests that doctors often use to evaluate a patient's dependence on alcohol are called the CAGE and Alcohol Use Disorders Identification Test (AUDIT) questionnaires. The CAGE questions are usually asked face to face in the doctor's office, but the AUDIT can be filled out with paper and pencil.

The CAGE questions:

- Have you ever felt the need to cut down on your drinking?
- Do people annoy you by criticizing your drinking?
- Have you ever felt guilty about your drinking?
- Do you ever need an eye-opener in the morning after a night of drinking?

The AUDIT questionnaire has ten questions:

• How often do you have a drink?

- How many drinks do you have on a typical day when you have a drink?
- How often do you have six or more drinks on a single occasion?
- How often during the past year have you found that you couldn't stop drinking?
- How often during the past year have you failed to do something you should have done because of drinking?
- How often during the past year have you needed a drink first thing in the morning?
- How often during the past year have you felt sad or guilty after drinking?
- How often during the past year have you been unable to remember what happened after drinking?
- Have you injured yourself or anyone else as a result of drinking?
- Has a friend, relative, or doctor expressed concern about your drinking?

known as fetal alcohol syndrome or FAS, affects one or two of every 1,000 babies born in the United States each year.

There is no single way that people become alcoholics. One reason why the experience of the disease is so difficult to describe is that the speed at which it develops and its consequences vary from person to person. Some people say that they became alcoholics after their first few drinks. Others drank responsibly for years and then became dependent on alcohol in midlife or even old age. In addition, some people get help before they get into serious trouble from drinking. They are sometimes called "high-bottom" alcoholics—that is, they "hit bottom" with their drinking while they still have a job or family. Alcoholics who have found themselves in trouble with the law, are homeless, or have lost their jobs and families sometimes refer to themselves as "low-bottom" alcoholics.

## Demographics

According to a study of adults in the United States conducted by the National Longitudinal Alcohol Epidemiologic Study, 44 percent of American adults are social drinkers; 22 percent drank at one time but do not drink alcohol at present; and 34 percent have always abstained from alcohol. Between 8 and 10 percent of adults are diagnosed with alcohol abuse or dependency in an average year.

Risk factors for alcoholism include:

- Genetics. Although no specific genes have been identified as causes of alcoholism, a study done in 2006 estimated that genetic factors account for 50 to 60 percent of a person's vulnerability to alcohol abuse.
- Family history of alcoholism.
- The age at which a person started to drink. People who began to drink before age sixteen are at higher risk of alcohol abuse.
- Gender. Men are two to three times more likely to become alcoholics than women. Women, however, do not metabolize alcohol as efficiently as men and can become intoxicated on smaller amounts of alcohol than men of the same weight.
- Race and ethnicity. Native Americans are at increased risk of alcohol abuse, followed by Hispanics, Caucasians, African Americans, and Asian Americans.
- A history of depression or other mental disorders.

## **Causes and Symptoms**

Alcoholism is the end result of a combination of factors that interact in ways that are unique to each person with the disorder:

- Genetic factors. These are thought to account for at least half of the cases.
- Psychological and emotional stress. Some people turn to alcohol to cope with anxiety, depression, posttraumatic stress disorder, or other painful feelings. This misuse of alcohol is called self-medication.
- Social and cultural factors. People may begin to drink heavily to look grownup or because their friends are encouraging them to drink. In some groups, heavy drinking may be seen as proof of masculinity. In addition, many films and television shows make drinking look glamorous or sophisticated.

• Body chemistry. Long-term drinking alters the levels of various chemicals in the brain, leading the person to crave alcohol either to get rid of bad feelings or to restore good feelings. Some people find that they eventually need alcohol just to feel normal.

The main symptom of alcoholism is denial; that is, alcoholics deny that they have a drinking problem until a series of health problems, family arguments, job losses, arrests, or other negative consequences force them to admit their drinking is out of control. Other symptoms commonly include:

- Drinking alone or in private.
- Hiding alcohol in the car, office, or other secret places.
- Feeling irritated or angry if unable to have a drink at the usual time.
- Gulping drinks quickly or drinking just to feel normal.
- Having problems with employment, finances, or the law.
- Building up tolerance to alcohol; that is, needing larger amounts to get the same effects.
- Having withdrawal symptoms after going without alcohol for a few days. These symptoms include shaking, insomnia, nausea, and vomiting.
- Feeling a need or compulsion to drink.
- Having blackouts. A blackout describes when someone is unable to remember what happened while he or she was drinking but did not pass out.

#### Diagnosis

The diagnosis of alcoholism is usually made on the basis of the patient's history and a review of his or her answers to screening questionnaires. The two screeners most often used are called the CAGE and AUDIT questionnaires (see sidebar). They can be completed in the doctor's office. People who answer yes to two of the four CAGE questions are seven times more likely to have a drinking problem than people in the general population. The AUDIT questionnaire assigns points ranging from zero to four depending on the frequency of the behaviors mentioned in the questions. A score of eight or higher indicates a high likelihood of alcoholism.

Although there are blood and urine tests that can detect the presence of alcohol or its breakdown products in the body, these tests reflect only whether the person was drinking at the time the test was given. Such tests are not useful in detecting long-term drinking patterns. In general, doctors often miss the diagnosis of alcoholism in their patients. This occurs because of the patient's denial or the patient's fear that the doctor will tell his or her employer or other family members.

#### Treatment

Treatment of alcoholism is important not only for the disease itself but also as a necessary first step toward treating other disorders. For example, psychiatrists will not treat someone for a mental disorder like bipolar disorder or depression as long as the person is still drinking. Patients with liver disease and other physical problems related to drinking also need to be treated for alcoholism before their physical health will improve.

Treatment of alcoholism usually proceeds in several stages.

- Intervention. Intervention is the term used for persuading the alcoholic to get help. It may be done by family members, the person's employer, a doctor, or others concerned about the person. The reason that intervention is usually necessary is that only 15 percent of alcoholics seek help on their own.
- Detoxification. Sometimes nicknamed detox, this is a process that takes between four and seven days. The alcoholic is placed in a treatment center where he or she is given medications (usually tranquilizers) to control the symptoms of withdrawal from alcohol. These symptoms may include sweating, nausea, vomiting, seizures, or a severe reaction called delirium tremens (also called the DTs). In delirium tremens, the person may hallucinate and have very high blood pressure and rapid breathing. It can be fatal if not treated.
- Rehabilitation. Rehabilitation for alcoholics includes patient education about the effects of alcohol, support groups, psychological counseling, and an emphasis on getting the patient to accept the truth about his or her alcohol dependence.
- Medications. There are several drugs, the best-known of which are Antabuse and Campral, which work either by making the person feel sick if he or she drinks or by lowering the craving for alcohol. These drugs work best in highly motivated patients. However, they are not magic cures for alcoholism.

Most doctors think that alcoholics should completely give up drinking because of the danger of relapse (falling back into out-of-control drinking). Although a few people who are not physically dependent on alcohol but have found themselves in trouble for abusing it can sometimes manage by simply cutting back on the amount they drink, complete abstinence is the best option for recovery.

### Prognosis

There is no cure for alcoholism. Recovery requires lifelong commitment to abstaining from alcohol and being honest with oneself. The prognosis is better for alcoholics who do not smoke and are not addicted to other drugs. Nonetheless, only about 30 percent of alcoholics maintain their recovery over the long term. Studies have shown that about half of those who complete a detoxification program relapse within six to twelve months.

## Prevention

People who have a family history of alcoholism can lower their risk of becoming alcoholics by learning about alcoholism and building a strong social support network. Parents can help by setting an example of responsible use of alcohol and talking openly about the physical and mental dangers of heavy drinking.

## The Future

According to statistics kept by the National Institutes of Health (NIH), the rate of alcoholism and alcohol abuse is rising in the United States and other developed countries. Some of the reasons for this trend are the easy availability of alcohol (including homemade alcoholic beverages); the role of the mass media in promoting drinking; and the weakening of family ties.

SEE ALSO Child abuse; Fetal alcohol syndrome; Smoking; Stroke

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Abstinence:	Complete stopping of alcohol	
consumpti	on.	

Blackout: Alcohol-related memory loss.

**Delirium tremens:** A severe physical reaction to withdrawal from alcohol in which the person hallucinates and has unstable blood pressure and breathing patterns.

**Detoxification:** A process or treatment program for clearing an alcoholic's body of alcohol. It

usually includes medications to help manage the physical symptoms of withdrawal.

**Relapse:** Returning to uncontrolled drinking.

- **Tolerance:** The need for greater and greater amounts of a drug to get the desired effects.
- Withdrawal: A group of physical and emotional symptoms associated with stopping the consumption of alcohol after long-term heavy use.

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## Definition

An allergy or allergic reaction is the body's response to an allergen, a foreign substance that enters or comes into contact with the body. Normally, the body's immune system produces antibodies to fight disease. In an allergy, the body overreacts to the allergen, which may be something rather harmless. Allergic reactions may be caused by the production of an antibody called immunoglobulin E (IgE).

### Description

The experience of an allergy depends on the body system or tissues that are affected, the severity of the reaction, and whether the substance that triggers the allergy can be eliminated from the patient's diet or living

#### Also Known As

Hypersensitivity reaction

#### Cause

Immune system's reaction to an allergen

#### Symptoms

Skin rash, sneezing, runny nose, watery eyes, nausea, diarrhea, headache

#### Duration

Hours to months

27

Allergies

situation. Some allergies affect only a small part of the body, like a skin rash on the wrist caused by touching certain metals in inexpensive costume jewelry. Other allergies may affect several major body systems, as in the severe reactions that some people have to bee stings or certain medications. These reactions are especially related to IgE antibodies. Some allergies are caused by substances that can be avoided, such as certain foods, while others may be triggered by exposure to plant or tree pollens that are hard to avoid when the plants are in season.

In general, when a person who has allergies comes into contact with an allergen, the person's immune system produces IgE antibodies that are specific to that allergen. The IgE antibodies trigger the release of a chemical called histamine. The histamine, in turn, causes the tissue swelling, difficult breathing, skin rash, watery eyes, runny nose, and other symptoms of an allergy.

The release of the histamine often attracts other cells from the immune system to the affected part of the body, increasing the inflam-

mation, tissue swelling, and production of large amounts of mucus otherwise making the patient's symptoms worse.

Researchers do not yet fully understand why some substances produce allergies in susceptible people and others do not, nor why some people do get allergies and others do not.

The most common types of allergies among people in North America are:

- Hay fever.
- Allergic conjunctivitis (inflammation of the tissue that lines the eyelid).
- Asthma.

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• Eczema. Eczema is sometimes called atopic dermatitis because it is a hypersensitivity reaction that affects a part of the body that is not in direct contact with the allergen. Hay fever and asthma are also considered atopic allergies. Boy with a rash that is an allergic reaction to the drug amoxicillin, an antibiotic. © PHOTOTAKE INC. / ALAMY.





*Man injecting himself with allergy medication.* SHUTTERSTOCK.

- Hives.
- Severe reactions to insect stings, certain foods, and certain medications. This type of severe allergic reaction is called anaphylaxis.
- Food allergies. Most cases of food allergies are caused by just eight foods: eggs, fish, milk, peanuts, shellfish, soy, tree nuts (walnuts, hazelnuts, almonds, etc.), and wheat.
- Contact dermatitis. In contrast to eczema, contact dermatitis is an inflammation of the skin caused by direct contact with an allergen or irritating substance, such as poison ivy, certain dyes, or certain metals, especially nickel.

## **Demographics**

Allergies are very common health problems in all age groups in the general population. About 50 million people in the United States suffer from some type of allergy, many of them from more than one. For example, about 20 percent of people who get hay fever eventually develop

asthma, and some develop eczema as well.

Some statistics for specific allergies are as follows:

- Hay fever: 20 percent of the population in developed countries.
- Asthma: 20 million people in the United States.
- Anaphylaxis: Affects between 1 and 3 percent of the population in developed countries.
- Eczema: about 15 million people in the United States.
- Hives: 20 percent of the population in developed countries.
- Food allergies: about 8 percent of school-age children.

There are two major types of risk factors for allergies: genetics and the environment. Allergies run in families, with allergic parents likely to have allergic children. Children are not necessarily allergic to the same substances as their parents. For example, the parent could be allergic to ragweed pollen while the child is allergic to peanuts. In general, allergies are more severe in children than adults. Several studies have shown that IgE levels are highest in young children and fall rapidly in people between the ages of ten and thirty years. Among children, boys are almost twice as likely as girls to develop allergies, but the sex difference disappears among adults.

Environmental factors are thought to explain why the rate of allergic diseases has increased around the world since the 1980s. These factors include dietary changes, a rise in air pollution, an increase in the levels of other allergens in the environment, and the fact that children are exposed to fewer infectious diseases than they were in the past. Some doctors think that exposure to and recovery from these diseases made children less susceptible to allergies.

## **Causes and Symptoms**

An allergic reaction is caused by the individual person's immune response to an allergen.

The symptoms of allergies may appear in one or more body systems:

- Skin: Redness, itching, bumps or wheals, crusting, weeping patches, blisters.
- Respiratory tract: Wheezing, sneezing, coughing, difficulty breathing, stuffy or runny nose.
- Digestive tract: Nausea, vomiting, diarrhea, stomach cramps.
- Eyes: Itching, watering, bloodshot or puffy appearance.
- Other: Headache, dizziness, ringing in the ears.

## Diagnosis

In some cases the allergen is easy to identify because the person has a pattern of symptoms that appears after exposure to the food, substance, or other trigger. In other cases the person may have to consult a specialist to identify the allergens that are causing the symptoms. The usual method of testing involves either a skin test, in which a small quantity of a possible allergen is injected under the skin, or a blood test known as the radioallergosorbent test (RAST). The RAST test is usually done in place of the skin test when the patient has had severe reactions to the allergen.

## Treatment

Anaphylaxis is a medical emergency that requires immediate treatment by a medical professional or emergency rescue team.

There is no cure for allergies. However, most can usually be treated with antihistamines and other medications that the doctor may recommend, depending on which part of the body is affected. The most important step in treatment, however, is avoiding the allergen if at all possible. Minimizing exposure to the allergen may involve installing air filters in the home, eliminating certain foods from the diet, cleaning the house frequently, moving to a part of the country with fewer pollen-producing plants, or other measures that the doctor may recommend.

Another approach to treating hay fever, asthma, allergic conjunctivitis, and some other allergies is desensitization, which is also called immunotherapy. In desensitization, patients are given a series of injections of their specific allergen under the skin, with the concentration of allergen in the shots being gradually increased. It takes an average of eight to twelve months for the patient to see results, however, and the injections must be taken for at least three years and sometimes closer to five years.

Although several alternative therapies—including herbal medicine, traditional Chinese medicine, and naturopathy—have been tried as treatments for allergies, a long-term study conducted by the Mayo Clinic in 2006 concluded that none of these was effective.

#### Prognosis

Most allergies are not life-threatening, the exceptions being anaphylaxis and asthma. About 5,500 people die each year in the United States from asthma and another 500 from anaphylaxis. In general, allergies tend to become less severe as people grow older. The major exception is food allergies (especially to peanuts), which can last a lifetime.

#### Prevention

There is no way to prevent people from developing allergies because the genetic factors that are involved in allergic reactions have not been fully understood. Desensitization therapy and avoidance of specific allergens are the only effective methods of prevention at present.

### The Future

Public health experts expect allergies of all types to become more common in the developed countries in the future as a result of lifestyle changes and a larger proportion of the world's population living in large cities. As of 2008, some researchers were working on a vaccine that would

## WORDS TO KNOW

- **Allergen:** Any substance that causes an allergic reaction in a person or animal.
- Anaphylaxis: A severe allergic reaction to a trigger (most commonly a food, medication, insect sting, or latex) that involves most major body systems.
- **Antibody:** A protein found in blood that is specific to a particular foreign substance, which may be an allergen or a disease organism. The antibody identifies that antigen and neutralizes it.
- **Atopy:** The medical term for an allergic hypersensitivity that affects parts of the body that are not in direct contact with an allergen. Hay fever, eczema, and asthma are all atopic diseases.
- **Contact dermatitis:** Inflammation of the skin caused by direct contact with an allergen or irritating substance, such as poison ivy, certain dyes, or certain metals.
- **Dander:** Tiny skin, feather, or fur particles from household pets that cause allergic reactions in some people.

- **Dermatitis:** The medical term for inflammation of the skin.
- **Desensitization:** A form of treatment for allergies that involves a series of shots containing the allergen to reduce the patient's sensitivity to that particular trigger. Desensitization is also called immunotherapy.
- **Histamine:** A chemical contained in mast cells that is released during an allergic reaction.
- Immunoglobulin E (IgE): An antibody in blood that activates mast cells during an allergic reaction.
- **Mast cells:** Specialized white blood cells that are found in connective tissue and contain histamine.
- **Wheal:** A suddenly formed flat-topped swelling of the skin; a welt.

target the IgE antibody itself. The vaccine is expected to be more effective than current desensitization therapy in preventing allergies.

**SEE ALSO** Anaphylaxis; Asthma; Conjunctivitis; Dermatitis; Eczema; Hay fever; Hives

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## Alzheimer Disease

## Definition

Alzheimer disease, or AD, is a terminal disease of the central nervous system that has no effective treatment or cure. Its most notable symptom is dementia—the loss of a person's memory and other cognitive functions beyond the effects of normal aging. It is named for Alois Alzheimer (1864–1915), a German doctor who first described a patient with the disease at a medical meeting in 1906.

## Description

AD affects a person's ability to think clearly, to remember, to speak, to pay attention, and to solve problems. The disease is characterized by the buildup of abnormal protein deposits in the brain, some of which look like clumps or plaques under the microscope and others that look like tangled fibers. Researchers do not yet know whether these abnormal proteins are causes of AD or whether they are a consequence of the disease.

## Also Known As

AD, Alzheimer dementia

#### **Cause** Unknown

#### Symptoms

Progressive loss of mental and body functions

**Duration** Five to 20 years



Illustration showing parts of the brain affected by Alzheimer disease. © NUCLEUS MEDICAL ART, INC. / ALAMY.

In addition, the cells in the part of the brain called the cerebral cortex begin to die and the tissue in that part of the brain begins to shrink. The cortex is the part of the brain that governs memory, the ability to pay attention, to use language, and to think; this helps to explain why these mental functions are gradually lost in patients with Alzheimer disease.

As the disease progresses, the destruction of brain tissue spreads to the areas of the brain that govern the emotions, and finally to the brain stem, which controls basic body functions like breathing and swallowing. Most patients with AD die from pneumonia or another infectious disease before the brain stem is completely destroyed.

There are two basic forms of AD, an early-onset form that affects people younger than 60 years of age and is transmitted within families; and a late-onset form that affects people over 60.

## **Demographics**

AD is presently the third leading cause of death in American adults, after cancer and heart disease. It is thought to affect 5 million adults in the United States as of the early 2000s—14 percent of the population over sixty-five. The frequency of Alzheimer disease increases with age; about 40 percent of people over eighty have AD. The lifetime risk of an individual's developing AD is between one chance in four and one chance in two.

## Famous People Who Had AD

Alzheimer disease can strike anyone in any walk of life; it has claimed the lives of a number of wellknown people:

- Dana Andrews, actor
- Rudolf Bing, director of the Metropolitan Opera
- Charles Bronson, actor
- Perry Como, singer
- Aaron Copland, composer
- Ralph Waldo Emerson, philosopher
- Geraldine Fitzgerald, actress
- Barry Goldwater, former Senator from Arizona
- Rita Hayworth, actress
- Charlton Heston, actor
- Willem de Kooning, painter
- Bill Mauldin, cartoonist
- Burgess Meredith, actor
- Iris Murdoch, novelist
- Otto Preminger, movie director
- William Proxmire, former Senator from Wisconsin
- Ronald Reagan, former President of the United States



#### Ralph Waldo Emerson.

- Sugar Ray Robinson, boxer
- Norman Rockwell, painter
- Jonathan Swift, writer
- E. B. White, writer
- Harold Wilson, former Prime Minister of Great Britain

Women are at greater risk than men of developing AD in old age, although the reasons for this difference are not clear. Race does not appear to be a risk factor for AD.

## **Causes and Symptoms**

The causes of AD are not fully understood. Early-onset AD, which is sometimes called familial Alzheimer disease, is caused by mutations of certain genes on chromosomes 1, 14, 19, and 21. This form of AD accounts for only 4–5 percent of cases, however. Most cases of Alzheimer's are considered sporadic, which means that they occur at random. Scientists think, however, that as many as 80 percent of these sporadic cases are influenced by genetic risk factors that have not yet been completely identified. Over 400 genes had been tested for links to AD as of 2007.

Other factors that have been identified as increasing a person's risk of developing AD include:

- High blood pressure or high blood cholesterol levels
- Heart disease or stroke
- Head injury
- Diabetes
- Female sex

Although some researchers in the 1970s thought that aluminum in drinking water, antiperspirant deodorants, and the diet might be a risk factor for AD, those theories have been disproved.

The symptoms of Alzheimer disease change as the disease progresses. Doctors generally describe four stages in the development of AD:

- Mild cognitive impairment. This phase can begin as long as eight years before a person is diagnosed with AD. The affected individual has some short-term memory loss and loses the ability to learn new information.
- Early dementia. The person begins to have language problems as well as memory problems, although they can still communicate with others. They may begin to need help with some activities of daily life. Many patients are diagnosed at this point because family and friends begin to notice changes in behavior or general loss of functioning.
- Moderate dementia. The person loses the ability to live independently and suffers further memory loss. Emotional control is lost; some patients with AD become physically violent toward others at this point as well as generally irritable or weepy. About 30 percent of patients in this stage have hallucinations and become unable to identify close relatives.
- Advanced dementia. The person loses the ability to talk except for occasional words, loses control of bowel and bladder functions, becomes unable to feed him or herself, and may be completely bedridden.

#### Diagnosis

There is no definitive test for AD except analyzing a sample of brain tissue, which can only be performed after the person dies. When a person first begins to show signs of dementia, however, doctors can arrive at a tentative diagnosis of AD through a series of tests and imaging studies. These may include:

- Laboratory tests of blood and urine samples. These tests are done to rule out infections or heart problems that might affect memory and other cognitive functions.
- Imaging studies, usually an MRI or a CT scan. These are done to rule out brain tumors, Parkinson disease, infections of the brain, or stroke.
- Mental status examination (MSE). This type of test evaluates the person's awareness of their location in time and place; their ability to follow simple instructions; their short-term memory; and their overall mood. The tests may include being asked to name the current president, to spell a word backward, or to copy a design on a piece of paper.
- A PiB PET scan. PiB stands for Pittsburgh Compound B, a chemical used in imaging tests which was shown in 2007 to identify the location of abnormal protein deposits in the brain of a patient with AD. This technique is not yet widely available in the United States, however.

### Treatment

There is no effective treatment for AD. There are three drugs that have been approved by the Food and Drug Administration (FDA) to slow down the loss of brain function in patients with AD, but these drugs work for only about six to twelve months, and benefit only half the people who take them. Patients who become depressed, agitated or violent can be given medications to help control these symptoms. There are no effective surgical treatments for Alzheimer disease.

Patients in the final stages of Alzheimer disease are kept clean and made as comfortable as possible. They may be given fluids and nutrients intravenously if they are unable to swallow. Death is most often caused because the patient stops breathing.

#### Prognosis

There is no cure for Alzheimer disease. Most people live between three and ten years after being diagnosed with the disorder, although some have lived as long as twenty years after diagnosis. Death usually comes as the result of pneumonia, a urinary tract infection, or complications following a fall.

### Prevention

There is no way to prevent the development of the familial form of AD in people who have the genes for the disease. With regard to sporadic lateonset AD, research is under way to identify steps that people can take to lower their risk of developing Alzheimer disease in old age. What researchers think as of the early 2000s is that the best way to protect the brain is to protect the heart: eating a healthful diet rich in fruits and vegetables, exercising regularly, getting treatment for high blood pressure, and keeping mentally active are all recommended. Such activities as playing chess, solving crossword puzzles, and social get-togethers with other people have been reported to delay the onset of AD or reduce its severity. Another suggestion is to take Advil or Motrin, as anti-inflammatory drugs may also lower the risk of AD.

#### The Future

Alzheimer disease is a major concern to policy makers in the twenty-first century as the proportion of elderly people in the general population continues to increase. Since the risk of developing AD doubles every five years after a person reaches 65, the possibility of having large numbers of people requiring several years of skilled nursing care at the end of their lives is worrisome to many doctors.

At present, many patients in the early and middle stages of the disease are cared for by relatives. The stress of caring for a parent, sibling, or spouse with AD is very high; many caregivers end up having to quit their jobs in order to care for the sick family member. In addition, the emotional strain of caring for someone who no longer recognizes the caregiver and may be openly violent or wander away from home can be exhausting. Doctors are increasingly aware that caregivers of Alzheimer patients have a high rate of depression and physical illnesses related to stress.

Research on AD is focused on preventive strategies. One possibility is developing a drug that can clear the abnormal proteins involved in

## WORDS TO KNOW

**Brain stem:** The lower part of the brain directly connected to the spinal cord. It controls such basic body functions as breathing and consciousness.

**Dementia:** Loss of memory and other mental functions related to thinking or problem-solving.

**Cerebral cortex:** The part of the brain that controls thinking, memory, paying attention, decision making, and using language.

AD from the brain tissue or prevent them from forming in the first place. Some experimental drugs of this type are currently undergoing clinical trials. Some researchers predict that there will be a major breakthrough in treating Alzheimer disease by 2020.

SEE ALSO Creutzfeldt-Jakob disease; Huntington disease

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## Anaphylaxis

### Definition

Anaphylaxis is a severe allergic reaction to a trigger (most commonly a food, medication, insect sting, or latex) that involves most major body systems.

## Description

Anaphylaxis is a reaction that occurs in people who are severely allergic to certain foods or other substances. People with individual or family histories of hay fever, asthma, eczema, or food allergies are at increased risk of having an anaphylactic reaction at some point in their lives. People with asthma are particularly likely to have an anaphylactic reaction to food at some point. Anaphylaxis is an overreaction to an allergen caused by the body's production of an antibody called immunoglobulin E or IgE. When a person who is susceptible to allergies first comes into contact with an allergen, the person's immune system produces IgE antibodies that are specific to that allergen. The IgE antibodies then attach themselves to the surface of mast cells, which are specialized cells that lie within the connective tissue near the blood vessels. The next time the person encounters that specific allergen, the IgE antibodies trigger the release of a compound called histamine from the mast cells. The histamine in turn causes the tissue swelling, difficult breathing, and other symptoms of anaphylaxis.

#### Also Known As

Immunologic reaction, anaphylactic shock

#### Cause

Severe allergic reaction

#### Symptoms

Difficulty breathing, nausea, vomiting, shock, loss of consciousness

#### Duration

Starts a few minutes to a few hours after exposure

#### Anaphylaxis

The EpiPen, a medicine that is used to treat severe allergic reactions that can result in anaphylaxis. © PAUL RAPSON / ALAMY.



Some people have a recurrence of symptoms about six to eight hours after an episode of anaphylaxis. This recurrence is called a biphasic or delayed reaction.

Some substances are more likely to trigger anaphylactic reactions than others. The most common triggers are:

- Foods. Almost any food can cause anaphylaxis in persons who are sensitive to it; however, peanuts, nuts from trees (walnuts, cashews, and Brazil nuts), shellfish, fish, milk, and eggs are the most common offenders.
- Stinging insects. Yellow jackets, honeybees, paper wasps, hornets, and fire ants are the most likely to cause anaphylactic reactions.
- Medications. The drugs most likely to cause problems include penicillin and other antibiotics; antiseizure medicines; fluids given after surgery; antibodies made from horse serum; blood and blood products; radiocontrast dyes used in certain types of imaging studies; and morphine or similar pain medications.
- Latex. Latex is a natural material made from the rubber tree that is often used to make surgical gloves and other medical products. The greatest danger of anaphylaxis caused by latex occurs when the rubber material touches moist areas of the body or internal surfaces during surgery, because the allergen in latex can be absorbed by the body much more quickly.

- Exercise. Some people develop anaphylaxis when they exercise within a few hours of eating certain foods.
- Other causes. Although such cases are rare, semen, hormones, and extreme temperatures have been reported to trigger anaphylaxis in some people.

## Demographics

The exact number of cases of anaphylaxis in the United States each year is unknown, partly because some doctors use the term to refer to relatively mild allergic reactions while others use it to describe only those severe enough to be considered life-threatening. About 30,000 people are taken to hospital emergency rooms with an anaphylactic reaction in an average year. A commonly cited statistic is that about 15 percent of the general population is at risk of anaphylaxis, with one percent of the population at specific risk of anaphylaxis caused by medications and another one percent allergic to latex. It is estimated that there are between 500 and 1,000 deaths from anaphylaxis each year in the United States; fifty of these deaths are caused by insect stings. People of all races, both sexes, and all age groups are equally at risk of anaphylaxis. Elderly people, however, are more likely

## First Aid for Anaphylaxis

There are several steps that friends or bystanders can take if they see someone having an episode of anaphylaxis:

- Call 911 at once for emergency help.
- Check the person to see whether he or she is carrying an EpiPen or similar device for emergency treatment of anaphylaxis. If so, administer the medicine at once.
- Have the person lie flat on his or her back and raise the feet above heart level.
- Try to keep the person calm, as moving unnecessarily or getting panicky can make the anaphylactic reaction worse.
- Cover the person with a blanket to keep warm but do not give him or her anything to drink.
- If the person is vomiting or bleeding from the mouth, gently turn him or her on one side to prevent choking.
- If possible, find out what triggered the reaction and tell the emergency helpers when they arrive.

than younger people to die from anaphylaxis because they often suffer from other diseases or medical conditions. Research conducted at Harvard in 2007 suggests that there may be geographical differences in the rates of anaphylaxis in the United States, with a higher proportion of cases in the Northeast and lower rates in the West and South. The researchers are not certain of the reasons for these differences, however.

## **Causes and Symptoms**

Anaphylaxis results from the release of histamine from mast cells in response to the presence of an allergen. Histamine causes the blood vessels

to dilate (expand), which lowers blood pressure. It also causes fluid to leak from the bloodstream into the tissues, which in turn causes the itchy skin swellings known as hives, and also lowers blood volume. These effects can lead to shock. Fluid can also leak into the air sacs in the lungs and make it difficult to breathe. Angioedema, or hivelike swelling of the tissues around the eyes or in the throat and mouth, can be severe enough to block the patient's airway.

Anaphylaxis is considered a whole-body allergic reaction because so many different body systems can be affected:

- Skin: hives, angioedema, redness, itching, rash
- Digestive tract: nausea, abdominal pain or cramping, vomiting, diarrhea, itchy mouth or throat
- Circulation: pale or bluish skin color, weak or rapid pulse, feeling dizzy or lightheaded, low blood pressure, loss of consciousness, shock
- Breathing: wheezing, shortness of breath, throat tightness, cough, difficulty talking, chest pain or tightness, nasal congestion, trouble swallowing
- Psychological: panic, feelings of doom, intense anxiety

#### Diagnosis

Diagnosis of anaphylaxis is based on the patient's symptoms in most cases, particularly if the reaction is sudden. In many cases it will be obvious either from the patient's description or from friends or bystanders what triggered the reaction. The doctor or emergency crew will usually take the patient's pulse, check breathing and blood pressure, and administer adrenaline (epinephrine), a hormone that counteracts the effects of histamine. The patient's heart rate will be monitored and basic life support given until they can be taken to the hospital.

#### Treatment

Treatment of anaphylaxis before the patient is taken to the hospital usually consists of checking to see whether the patient's airway is open and administering oxygen or otherwise helping the patient breathe. Intravenous fluids are given if the patient's blood pressure is low. Adrenaline and antihistamines are also given to help open the airway, relieve hives and other skin reactions, and restore normal blood pressure. Once in the hospital, the patient may be given further treatment, including emergency surgery to open the airway if he or she is still having trouble breathing. Steroid medications may be given in the hospital to lower the risk of a delayed or biphasic reaction, and the patient will usually be kept in the hospital for a few hours to make sure there will not be such a reaction after returning home.

#### Prognosis

The prognosis for anaphylaxis is very good if the person carries and uses an EpiPen or similar device, or receives prompt emergency treatment. In some cases, however, people die from low blood pressure, shock, or cardiac arrest. The elderly are at higher risk for death from anaphylaxis.

#### Prevention

People who have had a severe allergic reaction even once should consult an allergist for evaluation and advice about preventive measures. This precaution is particularly important if the patient does not know what caused the anaphylaxis. The allergist can determine the patient's triggers, prescribe an EpiPen or TwinJect for self-treatment, advise the patient about any medications that may increase the risk of anaphylaxis, and in some cases give the patient allergy shots to decrease sensitivity to insect stings or antibiotic medications. Other preventive measures that people can take include: Wear a MedicAlert or similar tag that identifies the allergy for bystanders or emergency rescuers. Learn to read food labels carefully and asking restaurant staff about food ingredients when eating out. Lower the risk of insect stings by wearing long-sleeved clothing and avoiding bright colors and perfumes that attract insects. Be careful when drinking sweetened beverages outdoors. Avoid walking barefoot in the grass. If stinging insects appear, stay calm and avoid slapping or swatting at them; move away from them slowly.

#### The Future

Experts believe that the number of anaphylactic reactions in the United States will rise in the future because of the increased number of potential allergens to which people are exposed.

SEE ALSO Asthma; Eczema; Hay fever; Hives

## WORDS TO KNOW

- **Adrenaline:** A hormone that can be used in medicine to open the breathing passages in patients with severe angioedema. It is also called epinephrine.
- Allergen: Any substance that can provoke an allergic reaction in susceptible individuals.
- **Angioedema:** The medical term for the swelling of tissues around the eyes, lips, and genitals that sometimes accompanies hives.

- **Biphasic reaction:** A recurrence of the symptoms of anaphylaxis about six to eight hours after the first episode.
- **Histamine:** A compound contained in mast cells that is released during an allergic reaction.
- **Mast cells:** Specialized white blood cells that are found in connective tissue and contain histamine.
- **Shock:** A medical emergency in which there is a drop in blood pressure and a reduced volume of blood circulating in the body.

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# m

## Anemias

The anemias are a group of blood disorders characterized by too little hemoglobin in the blood. Hemoglobin is a substance contained in red blood cells that carries oxygen from the lungs to other body tissues. Anemia is often a sign or symptom of an underlying disease rather than a disease in its own right. There are three tests commonly used to detect anemia: the number of red blood cells can be counted; the amount of hemoglobin in the red blood cells can be measured; or the proportion of blood cells to serum (the liquid part of blood, called the hematocrit) can be assessed.

Anemia can develop in three ways: loss of blood through injury, diseases of the digestive tract, or heavy menstrual flow in women; rapid destruction of red blood cells (for example in sickle cell anemia); or inadequate production of healthy red blood cells (for example in thalassemia). The underlying causes of anemias range from poor nutrition (iron-deficiency anemia) and digestive disorders (Crohn disease, celiac disease) to colorectal cancer, parasitic diseases (hookworm), and genetic disorders (sickle cell anemia, thalassemia).

The anemias are the most common type of blood disorder in the general population, affecting 4 percent of men and 8 percent of women in the developed countries, and two to five times as many people in the poorer nations.

**SEE ALSO** Celiac disease; Colorectal cancer; Crohn disease; Heart failure; Lead poisoning; Prematurity; Restless legs syndrome; Sickle cell anemia; Thalassemia; Ulcers



## Definition

Anorexia is an eating disorder—a psychiatric condition marked by fear of gaining weight, a distorted mental picture of what one's body actually looks like, and extreme dieting in order to lose weight. Many anorexics also exercise for long periods or take diuretics (drugs that increase urine output) in order to lose weight.

#### Description

The *Diagnostic and Statistical Manual of Mental Disorders* fourth edition (DSM-IV), the reference that doctors use to diagnose mental illness, specifies that a person must have a body weight that is only 85 percent of normal for their sex, age, and height, combined with a fear of becoming fat and a preoccupation with the size and shape of the body in order to be diagnosed with anorexia. In addition, a girl must miss three or more menstrual periods in a row to fit the diagnostic criteria.

Other signs that are commonly seen in young people with anorexia include osteoporosis (brittle bones), slowed growth, hair loss, aches in the joints and muscles, feeling cold much of the time, low blood pressure, slowed heart rate, and dry skin.

#### Demographics

There are between eight and thirteen young people per 100,000 in the United States and Canada who meet the strict DSM-IV criteria for anorexia. On the other hand, many doctors think there are other adolescents and young adults who have a milder form of anorexia but are not counted because they do not meet all the DSM-IV criteria; thus the disorder may be more widespread than the official figures indicate.

Anorexia is most common in teenagers and young adults; 40 percent of diagnosed patients are between fifteen and nineteen years old. A few patients begin to show signs of the disorder between the ages of seven and twelve. At the upper end of the age spectrum, 10 percent of patients are women over the age of forty; it is estimated that between 1 and 3 million women in the so-called baby boomer generation are struggling with eating disorders. The

#### Also Known As Anorexia nervosa, AN

#### Cause

Social, genetic, and psychological factors

#### Symptoms

Severe dieting and exercise, preoccupation with weight and body shape

#### Duration

Appears in adolescence or early adulthood; may last for years if untreated female/male ratio for anorexia is thought to be nine females to every male. Although gay men are at somewhat higher risk of anorexia, heterosexual men can also develop the disorder.

At one time it was thought that anorexia is primarily a disease of middle-class Caucasians; however, in recent years, more cases of the disorder are appearing in African American and Hispanic women.

### **Causes and Symptoms**

Although there is general agreement that anorexia is caused by a combination of factors, doctors disagree about which are the most important. There is considerable evidence that genetic factors play an important role. Another factor that is being researched is the role of a brain chemical involved in both anorexia and depression.

Other factors that have been cited by researchers include personality traits, family issues, and cultural values. Many anorexics are high achievers with a tendency toward perfectionism, and controlling their weight appears to be part of

a larger need to be in control of their lives. In terms of family issues, some psychiatrists think that anorexia may be a way for the young person to separate from their family and establish their own identity.

In addition, as many as 50 percent of girls hospitalized for anorexia were sexually abused as children, leading doctors to think that they may be afraid of growing into mature women and want to keep a childish figure as long as possible. Last, cultural emphasis on thinness as a mark of social status has been blamed for encouraging girls (and older women) to lose more weight than is healthful.

The symptoms of anorexia include psychological as well as physical symptoms. Anorexia affects most of the body's major organ systems:

• Heart and blood: abnormally slow heartbeat, low blood pressure, episodes of weakness and dizziness, low levels of calcium and magnesium in the blood, and anemia (red blood cell count that is too low)



Anorexics have a distorted view of their own bodies, seeing themselves as being much bigger than they really are. © TED FOXX / ALAMY.
# Anorexia and Athletes

Athletes are a high-risk group for anorexia, particularly in sports in which weight is important in competition. Runners, wrestlers, gymnasts, figure skaters, racing jockeys, and dancers, male as well as female, have an increased risk of developing anorexia and its associated problems of brittle bones and digestive disorders in later life. Women in sports are at risk of what is called female athlete triad, a group of three symptoms and signs that appear when a woman diets too strictly for competition: amenorrhea (stopping of menstrual periods); an unusual number of stress fractures; and an eating disorder, most commonly anorexia.

Many people also do not realize that anorexia can be literally fatal to women in sports. Several wellregarded athletes and dancers, such as Christy Henrich (1972–1994) and Heidi Guenther (1975– 1997), died in their early 20s of organ failure resulting from anorexia; Henrich, a gymnast, weighed only 47 pounds (21.3 kilograms) when she died.

- Muscles, bones, and skin: dry skin, loss of scalp hair, sore muscles, swelling of the joints caused by abnormal eating patterns, risk of developing osteoporosis, a bone disorder in which the bones break or fracture more easily than is normal as the result of the loss of minerals in bone caused by starvation
- Digestive system: constipation, stomach ulcers, bleeding in the digestive tract, and an increased risk of disorders of the liver and pancreas
- Temperature regulation: feeling cold, as the loss of body fat means that the body loses heat more rapidly
- Psychological symptoms: depression, distorted notions of the body, and an increased risk of suicide

### Diagnosis

Anorexia is usually diagnosed during an office visit to the patient's primary care doctor, although she or he may be sent to a psychiatrist for an additional evaluation. Primary care doc-

tors are now encouraged to give a screening test to an adolescent or young adult who seems unusually concerned about weight or asks the doctor a lot of questions about weight loss. These screeners are short sets of five questions about eating habits that the patient can quickly answer. Other questionnaires that the doctor may use include the Clinical Eating Disorder Rating Instrument (CEDRI) or the Eating Disorder Examination (EDE); these take a little longer for the patient to fill out.

If the doctor thinks that the patient may have anorexia, he or she can look for some of the physical signs that accompany the disorder after weighing the patient. Besides extreme thinness, these signs typically include a yellowish discoloration of the skin, signs of dehydration, an abnormally low blood pressure reading, and a slow heartbeat. The doctor will order laboratory tests of the patient's blood and urine to see whether the blood chemistry and the red blood cell count are normal. The patient will also be given an electrocardiogram (ECG) to check for abnormal heart rhythms and other potential heart problems.

#### Treatment

The treatment of anorexia is difficult and complicated, partly because many patients deny that they have a problem. It is common, in fact, for anorexics to argue with the doctor that they simply have a different lifestyle and should be left alone. Hospitalization may be necessary if the patient is having serious medical complications related to starvation or has threatened suicide. In the hospital she (or he) will be given fluids and nutrients intravenously at first in order to stop the weight loss and bring other physical problems under control. The patient will then be given psychotherapy (usually cognitive-behavioral therapy or CBT) and education about nutrition. Antidepressants may be given to treat anxiety or emotional depression, but they are not considered effective in treating the eating disorder by itself.

Anorexics who do not need to be hospitalized for serious physical conditions may be treated on an outpatient basis or in a day hospital. They are often referred to a dietitian for personalized advice about healthful eating patterns and sample menus. Group psychotherapy is considered less effective than either individual treatment or family therapy in treating anorexia because the patients in a treatment group are likely to compete to see who can stay the thinnest. Family therapy is particularly recommended for patients under eighteen and for families in which the mother has an eating disorder.

#### Prognosis

Anorexia is a potentially life-threatening illness with one of the highest death rates among psychiatric disorders; between 6 and 20 percent of patients die either from starvation or suicide. Because treatment is complicated and may take years, the prognosis varies. Most patients need monitoring over a period of months to years to help them maintain a healthy weight and not slip back into disordered eating habits. One very real problem since the coming of the Internet is the rise of a number of websites promoting anorexia, encouraging people with the disorder to consider it a valid lifestyle decision rather than an illness.

#### Prevention

There is not likely to be any new medical or psychological treatment for anorexia in the next few years. For now, prevention of anorexia depends

# WORDS TO KNOW

- **Amenorrhea:** Stopping of normal menstrual periods.
- Antidepressant: A type of drug given to treat eating disorders as well as mood disorders like anxiety and depression.
- **Cognitive-behavioral therapy (CBT):** An approach to therapy that aims at changing distorted thinking patterns, beliefs, and behaviors in order to change the patient's feelings.
- **Diuretic:** A type of drug that increases the body's production of urine. Some people with anorexia take diuretics to lose weight by reducing the amount of water in the body.
- Female athlete triad: A group of three symptoms that often occur together in female athletes: amenorrhea, osteoporosis, and disordered eating.
- **Osteoporosis:** A disease in which bones lose their density and are more likely to break or fracture under stress.

on early recognition of an unhealthy concern with weight, since girls as young as five or six are known to worry about their weight, especially if their mothers or older sisters are dieting. In some cases the child's pediatrician can help detect the problem or offer advice.

#### The Future

It can be difficult to change an entire society's obsession with thinness and a very limited notion of physical attractiveness. In addition, the availability of Web sites promoting anorexia increases the difficulty of teaching young people healthy eating habits even if they do not measure up to celebrity-inspired notions of glamour or beauty. A 2006 study at Stanford University found that 36 percent of the patients in the eating disorders unit of the university's hospital visited these Web sites, and of this percentage, 96 percent had learned new techniques for dieting or losing weight from the Web sites.

**SEE ALSO** Bulimia; Osteoporosis

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# Anthrax

# Definition

Anthrax is a disease caused by a rod-shaped bacterium called *Bacillus anthracis*. The bacterium forms spores (tough dormant forms of the organism) that can live for decades in the soil. Robert Koch (1843–1910), the German doctor who is considered the father of microbiology, identified the bacteria as the cause of anthrax in 1877.

Anthrax is primarily a disease of grazing animals such as sheep, goats, camels, and cattle. It rarely affects household pets like cats and dogs. It is one of the oldest animal diseases known to humans and is probably one of the ten plagues of Egypt described in the Book of Exodus in the Old Testament of the Bible. Animals can become infected by anthrax through

#### Anthrax

Anthrax sore on the skin. © MEDICAL-ON-LINE / ALAMY.



#### Also Known As

Woolsorter's disease, ragpicker's disease

#### **Cause** Bacteria

Symptoms (cutaneous) Black-centered sore on skin

#### Symptoms (intestinal) Nausea, vomiting, bloody diarrhea

#### Symptoms (inhalation)

Flu-like symptoms, high fever, difficulty breathing, shock

Duration (cutaneous) Three weeks

**Duration (intestinal)** Four to seven days

**Duration (inhalation)** Two to four days eating grass coated with anthrax spores or getting the spores into open wounds on their hides. Thousands of cattle in Europe and North America died each year from anthrax until French scientist Louis Pasteur (1822–1895) developed an effective vaccine against the bacteria in 1881.

### Description

Anthrax can take three different forms in humans. The most common form, accounting for 95 percent of cases, is cutaneous (skin-related) anthrax. It occurs when anthrax spores enter the body through a cut or break in the skin, producing a painless sore with a black center. Cutaneous anthrax is easily treated with antibiotics and is rarely fatal unless the infection spreads into the bloodstream.

The second form is gastrointestinal anthrax. Humans can get it by eating raw or undercooked meat from an infected animal. In this form of anthrax, the bacteria cause inflammation of the intestines leading to ulcers in the tissues lining the digestive tract. The infected person experiences nausea, vomiting, and bloody diarrhea. Between 25 and 60 percent of people with this form of anthrax die.

Inhalation anthrax is the third form, caused when anthrax spores enter a person's nose and throat and are carried to the lungs. The spores enter the lymphatic system and the lymph nodes in the central chest cavity. There, the spores germinate into active bacteria. The bacteria enter the bloodstream and are carried throughout the body. The bacteria then produce toxins that cause bleeding, tissue destruction, and eventual death.

Inhalation anthrax is fatal in about 75 percent of cases even when treated. This form of anthrax used to be called woolsorter's or ragpicker's disease because it was most likely to affect weavers and other people who worked with wool or hair taken from infected animals. Inhalation anthrax is the form most feared today because it is the form that develops when anthrax spores are used as a method of bioterrorism.

# Demographics

Anthrax is a relatively rare disease in humans in developed countries since the introduction of Pasteur's vaccine made it possible to protect animals from the disease. Other safety measures include the sterilization of animal waste materials.

Prior to the 2001 anthrax mailings, the last case of inhalation anthrax in the United States took place in 1976, when a weaver in California died after using wool imported from Pakistan. In 2006 an artist in the United Kingdom died from inhalation anthrax linked to working with untreated animal skins. The last known case of gastrointestinal anthrax in the United States occurred in 2000, when several people in Minnesota fell ill after eating meat from an infected steer.

#### 2001 Anthrax Mailings

Within a week of the terrorist attacks of September 11, 2001 on the United States, letters containing spores of a particularly powerful strain of the anthrax bacteria were mailed to several news offices and two U.S. senators. An expert molecular biologist who examined the spores in the letters described them as "weapons grade." Twenty-two people are known to have developed anthrax from the letters; half experienced the inhalation form of the disease. Five of the victims died: an editor at a Florida newspaper, a Vietnamese immigrant who worked in a New York hospital, an elderly widow in Connecticut, and two postal employees in Washington, D.C.

As of early fall 2008 the identity of the perpetrator is still uncertain. Suspicion was first directed toward a virologist named Steven Hatfill in 2003, then redirected toward a researcher at the Army medical research institute in Maryland named Bruce Ivins. Ivins killed himself at the end of July 2008 when he was informed that he was about to be prosecuted for the 2001 anthrax mailings. Although a team of scientists working since 2001 traced the anthrax samples in the letters to one specific flask of anthrax in Ivins's laboratory, some people think that someone other than Ivins may have had access to the flask and mailed the contaminated letters. The motive of the anthrax mailer as well as his or her identity may never be known.

People of all races and ages are equally likely to get inhalation anthrax when the bacillus is used as a weapon of bioterrorism. Cutaneous and gastrointestinal anthrax are rare in the United States. Young adults are the age group most commonly affected due to occupational exposure. People in the following occupations are at some risk of cutaneous anthrax:

- Veterinarians
- Farmers and ranchers

- · Forest rangers, field biologists, and others who study wildlife
- People who work with wool, animal hides, hair, or bone meal products

# **Causes and Symptoms**

The cause of anthrax is a bacterium, *B. anthracis*. The bacteria cause organ destruction and internal bleeding in animals and humans through the release of toxins that target the tissues lining blood vessels, lymph vessels, and the intestines. One of the toxins causes fluid to build up in the tissues and damages the body's immune system, while the other toxin kills infected cells directly.

It is important to understand that humans cannot transmit anthrax directly to one another, although a person's skin, hair, or clothing can be contaminated by anthrax spores. People can be decontaminated after exposure to anthrax spores by showering in hot water and using an antimicrobial soap. Suspected articles of clothing should be boiled in water for a minimum of thirty minutes or else burned. It is not necessary to isolate or quarantine a living patient diagnosed with anthrax after he or she has been decontaminated.

The symptoms of anthrax depend on the type:

- Cutaneous: The incubation period is between two and five days after the spores get into a cut or break in the skin. An itchy papule (raised skin lesion) appears, followed by a blackish painless ulcer about an inch across with a round, swollen edge. Nearby lymph nodes may be swollen. The ulcer lasts for about two weeks before separating from the skin and leaving a permanent scar. The infection spreads into the bloodstream in about 5 to 10 percent of untreated patients.
- Gastrointestinal: Symptoms appear between two and five days after eating infected meat. The patient experiences nausea, vomiting, painful abdominal cramps, loss of appetite, vomiting blood, and bloody diarrhea.
- Inhalation: The symptoms begin abruptly, usually within one to three days after exposure. However, in some cases it may take as long as forty-two days after exposure (and perhaps longer) for the person to feel sick. The person first notices coughing and mild discomfort around the breastbone, quickly followed by high fever, severe shortness of breath, coughing or vomiting blood, heavy sweating, and chest pain severe enough to be mistaken for a heart attack.

# Diagnosis

There is no screening test for anthrax. Diagnostic tests for the various forms of anthrax are ordered according to the patient's history—including his or her occupational history—and specific symptoms:

- Skin scraping or biopsy: A sample of tissue fluid from the characteristic skin ulcer caused by cutaneous anthrax can be stained and viewed under a microscope for evidence of the anthrax bacteria.
- Sputum culture. The secretions from the lungs of a patient with inhalation anthrax can also be examined under a microscope.
- Stool sample. Stool can be tested for anthrax bacteria in cases of suspected gastrointestinal anthrax.
- Endoscopy. An endoscope is a lighted flexible tube that can be inserted into the throat or the intestines to look for the characteristic ulcers caused by the anthrax bacteria.
- Chest x ray or computed tomography (CT) scan. These imaging studies can be done to look for signs of inhalation anthrax.
- Blood test and laboratory culture. This test can be done to check for any of the three forms of anthrax. The sample is cultured on a material called blood agar, which is made from animal blood combined with a gelatin-like substance derived from seaweed. If *B. anthracis* is present, the red blood cells in the gelatin surrounding the bacterial colony will be destroyed.

# Treatment

All three forms of anthrax are treated with oral or intravenous antibiotics. Penicillin is usually effective for cutaneous anthrax. However, ciprofloxacin and doxycycline are usually recommended for the other forms of the disease. Inhalation anthrax requires hospitalization and treatment with intravenous antibiotics. People who may have been exposed to anthrax may be given antibiotics for as long as sixty days to prevent inhalation anthrax.

# Prognosis

Cutaneous anthrax has the best prognosis; most people recover without complications except for a scar where the ulcer appeared. Gastrointestinal anthrax has a mortality rate of 25 to 60 percent, while inhalation anthrax has a death rate between 45 and 75 percent even when treated.

### Prevention

Anthrax can be prevented in several ways. One preventive measure is to give a sixty-day course of antibiotics to anyone exposed to the disease. Another preventive measure is to avoid eating raw or undercooked meat.

There is a vaccine against anthrax that was approved by the Food and Drug Administration (FDA) in 1970, but it is not given to the general public. Those who are eligible to receive the vaccine include:

- Active-duty military personnel in areas that carry a high risk of exposure to anthrax.
- Researchers and laboratory assistants who work with anthrax.
- People who must work with animal products in parts of the world where livestock are not routinely vaccinated against anthrax.
- People who work with animal hides or wool imported from these countries.

People or animals who die from anthrax must be buried or otherwise disposed of carefully in order to prevent contaminating others. The body of a person who is known to have died from anthrax is considered a biohazard because the skin and any body fluids can contaminate healthcare workers. The body must be placed in an airtight bag and preferably cremated, as burial will not kill anthrax spores.

## The Future

Anthrax is a major concern to public health officials because of its potential as an agent of bioterrorism. The Centers for Disease Control and Prevention (CDC) classifies anthrax as a Category A agent, the highest of three levels. Category A agents are defined as disease organisms that pose the greatest possible threat to public health; can be easily spread over a large geographic area; and require a high level of planning in order to protect the public. In addition, the fact that anthrax spores can survive in the environment for years makes the disease even more dangerous. In 1942, during World War II, the British deliberately tested anthrax as a bioweapon on a small island off the coast of Scotland. It took until 1990 for the island to be completely decontaminated.

SEE ALSO Ebola and Marburg hemorrhagic fevers; Plague; Smallpox

# WORDS TO KNOW

**Bioterrorism:** The use of disease agents to frighten or attack civilians.

Cutaneous: Pertaining to the skin.

- **Papule:** A small cone-shaped pimple or elevation of the skin.
- **Quarantine:** The practice of isolating people with a contagious disease for a period of time to prevent the spread of the disease.
- **Spore:** A dormant form of the anthrax bacteria that can live for decades before being reactivated and reproducing.

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# **O** Appendicitis

# Definition

Appendicitis is a condition in which the appendix, a finger-shaped projection connected to the large intestine, becomes inflamed and its opening blocked. It is a medical emergency.

# Description

Appendicitis develops when the opening of the appendix is blocked. This blockage can be caused by a number of different objects ranging from intestinal parasites to fecal matter, or by an infection. As the blockage progresses, the appendix begins to die from lack of blood flow. It is then invaded by bacteria and forms pus. If the condition is not treated, the appendix swells and eventually bursts, spreading the infection throughout the abdomen. This spread of infection and inflammation to the tissues lining the abdomen is called peritonitis and is a very dangerous condition. The pain of appendicitis usually starts two to three days before the appendix gets to the point of bursting. The person typically notices a vague discomfort in the area underneath the navel. Over the next day the pain gets worse and moves downward toward the lower right portion of the abdomen, near the right hip. The "classic" symptoms of appendicitis at this point are nausea, vomiting, low-grade fever, and loss of appetite. Fewer than 50 percent of patients with appendicitis, however, have the full set of classic symptoms. Children and the elderly are often misdiagnosed because they have fewer of these symptoms. As a result, their treatment can be delayed. The appendix ruptures before surgery in about 270 out of every 1,000 cases, and the rate of rupture is higher in children, pregnant women, and older adults.

# Demographics

The National Institutes of Health (NIH) estimates that about 7 percent of the general population in the United States will develop appendicitis at some point in life. There are about 1.1 cases per 1,000 people each year. The disorder is most common in people between the ages of ten and thirty, but it can develop at any age. In a few cases, appendicitis has been

#### Also Known As Epityphlitis

#### **Cause** Blockage and inflammation of the appendix

#### Symptoms

Severe pain in abdomen, loss of appetite, nausea, vomiting, fever, diarrhea or constipation

#### Duration

Four to 48 hours



*Illustration of appendicitis.* © PHOTOTAKE INC. / ALAMY.

diagnosed in newborn babies. Appendicitis is equally common in persons of all races and ethnic groups. It is slightly more common in men than in women, however. Although appendicitis is not hereditary, it does appear to be more common in some families.

# **Causes and Symptoms**

The basic cause of appendicitis is inflammation of the appendix resulting from an obstruction of some kind or an infection. The appendix can be blocked by an overgrowth of lymphoid tissue, food wastes, small pieces of hardened stool, worms or other parasites, foreign objects, or a cancerous tumor. It may also become inflamed as a result of trauma or infection, or as a complication of Crohn disease. The blocked appendix swells up with mucus, shutting down the blood vessels that supply it with blood. As its tissues die, bacteria from the intestine grow rapidly within it. If the infection is not stopped by surgical removal of the organ, the appendix will eventually burst and the bacteria inside it will spread to other parts of the abdomen. There is no single symptom that is unique to appendicitis, nor is there a "typical" group of symptoms that all

# Wartime Appendectomy on a Submarine

A remarkable story of battlefield medicine during World War II is an emergency appendectomy performed on a 19-year-old sailor aboard a submerged submarine, the USS Seadragon, in September 1942. There was no time or opportunity to take the sick sailor to a hospital on land and no qualified surgeons on the submarine. As the sailor got worse, the commanding officer explained that a pharmacist's mate might be able to perform an appendectomy. The sailor said that whatever the pharmacist's mate could do to help him was fine. The operation was performed on a table in the dining area with ether as the anesthetic and improvised instruments sterilized in boiling water. The pharmacist's mate removed the appendix, cleansed the stump with alcohol from a torpedo, and used crushed sulfa tablets to prevent infection, as no penicillin was available in 1942. Luckily, the sailor's 3-inch (7.6-centimeter) incision healed easily; he was back on duty in a few days. Speaking to a historian in 1999, the pharmacist's mate recalled the operation: "What was important was that I did my job.... It was my job to do anything I could to preserve life and, really, I didn't deserve special credit or recognition for doing that...."

patients experience. The following are the most common symptoms and the percentages of patients who report having them:

- Pain in the abdomen moving from the navel to the right lower part of the abdomen: 80 percent.
- Nausea: 85 percent.
- Fever: 60 percent.
- Loss of appetite: 74 percent.
- Diarrhea or constipation: 18 percent.
- Symptoms lasting less than 48 hours: 80 percent. About 2 percent of patients, however, report pain in the abdomen lasting as long as two weeks.
- A previous history of pain in the abdomen: 23 percent.

# Diagnosis

Diagnosis of appendicitis is tricky and complicated, partly because there are many diseases and disorders—particularly complications of pregnancy and Crohn disease—that cause abdominal pain, fever, and vomiting; and partly because at least half of all patients who have appendicitis do not have the classic symptoms of the condition. In addition, the size and location of the appendix varies somewhat. In

some patients the appendix is located on the left side of the body rather than the right, and in others the appendix is unusually long and extends from the right side toward the left side. Diagnosis of appendicitis is based on information from several different types of examinations and tests.

• Physical examination: The doctor will take the patient's history, record their temperature, and perform an examination of the abdomen. Patients with appendicitis typically feel what is called rebound tenderness (soreness) when the doctor first presses on the abdomen and then releases the pressure. The patient may also stiffen the muscles of the abdomen in response to pressure; this

reaction is called guarding. In addition, the doctor may be able to feel that the abdomen itself is rigid. Last, the doctor may move or rotate the patient's right leg or hip in order to test for pain during these maneuvers.

- Blood test: A high white blood cell count indicates the presence of infection.
- Imaging tests: These may include x rays, ultrasound, or computed tomography (CT) scans. The CT scan is the most commonly used imaging test to diagnose appendicitis, but x-ray studies can be useful for detecting foreign bodies or hardened stools that may be blocking the appendix.
- Urine test: This test may be done to rule out kidney stones or a urinary tract infection, but can be abnormal if the appendix inflammation is close to the urinary tract.

In a few cases involving women who may have a disorder of the ovaries or the fallopian tubes, the doctor may need to perform an exploratory type of surgery called a laparoscopy to see which organ is causing the patient's symptoms.

## Treatment

In a few cases, if the doctor is not certain of the diagnosis, he or she may prescribe a course of antibiotics to see whether the patient's symptoms are caused by something other than an inflamed appendix and may not require surgery. For most patients, however, surgery is the only cure for appendicitis. The surgeon can perform an appendectomy (surgical removal of the appendix) in several different ways. The oldest procedure is called an open appendectomy. The surgeon makes an incision (cut) between 2 and 4 inches (5.1 and 10.2 centimeters) and in length on the lower right side of the abdomen. The appendix is removed from its location and the area is rinsed with sterile fluid to prevent further infection. A newer and more commonly used technique is called a laparoscopic appendectomy. It requires much smaller incisions, only an inch (2.5 centimeters) or so long. The surgeon inserts a laparoscope, which is an instrument that allows the surgeon to see inside the abdomen, through one incision, and surgical instruments to remove the appendix through another small incision. If the surgeon finds that the infection has spread or that there are other complications, the operation may have to be completed as an open appendectomy. In March 2008, surgeons at a medical

# WORDS TO KNOW

Appendectomy: Surgical removal of the appendix.

- **Guarding:** Stiffening of the muscles in response to a doctor's touch.
- **Laparoscope:** A fiberoptic instrument resembling a telescope that can be inserted through a small incision to allow a doctor to see the inside of the abdomen during surgery.
- **Peritonitis:** Inflammation of the membrane that lines the abdominal cavity and covers some of the internal organs.
- **Pus:** A whitish-yellow material produced by the body in response to a bacterial infection. It consists of tissue fluid and dead white blood cells.
- **Rebound tenderness:** Pain experienced when the doctor releases pressure on the abdomen.

center in California removed a woman's appendix through her vagina. The procedure is still considered experimental but may allow female patients to recover more rapidly.

#### Prognosis

Most people do very well after an appendectomy if their appendix was removed before it ruptured. The average hospital stay is between one and three days after surgery, but full recovery at home may take a few weeks before the patient can return to vigorous exercise or lifting heavy objects. The mortality rate for appendicitis in the United States is very low, between 0.2 and 0.8 percent of patients; most of these deaths are caused by complications of peritonitis rather than by the appendectomy itself. The rate of complications in appendicitis increases tenfold if the appendix bursts before surgery.

#### Prevention

There is some evidence that people from cultures whose diets have a high level of fiber (the part of plants that is not digested) are less likely to develop appendicitis than those whose diets are low in fiber. It is thought that higher levels of fiber in the diet help the intestines push food along more efficiently, thus lowering the likelihood that the appendix will become blocked by fecal matter. Apart from increasing the amount of fiber in one's diet, however, there is no way to predict or prevent appendicitis.

## The Future

As of 2008, about 10 percent of patients with appendicitis were not correctly diagnosed on their first visit to the doctor. Ongoing research may help doctors lower the rate of missed diagnoses of appendicitis as well as develop improved surgical instruments and techniques.

#### SEE ALSO Crohn disease

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# m

# Arthritis

Arthritis is a group of disorders that affect the joints, causing pain, welling, and gradual loss of the ability to move freely or easily. About forty-six million Americans have some form of arthritis, and nineteen million have their activities limited by the disease. Osteoarthritis, the most common form, usually results from the aging process, infection of the joints, or repeated injuries to the joints; it is the leading cause of disability in adults over the age of fifty-five. The other common form, rheumatoid arthritis, is a disorder of the immune system in which the body attacks its own tissues. It has a much greater impact on the patient's life span and quality of life than osteoarthritis.

Arthritis can also develop in people diagnosed with such other diseases as lupus, Lyme disease, hepatitis, Crohn disease, or ulcerative colitis.

There is no cure for either osteoarthritis or rheumatoid arthritis. Treatment for these joint disorders is aimed at relieving pain and swelling, and keeping the affected joints functional. In the case of rheumatoid arthritis, treatment should begin as soon as possible after diagnosis to slow down the damage to the patient's joints.

SEE ALSO Osteoarthritis; Rheumatoid arthritis; Sprains and strains

# u

# Asperger Syndrome

# Definition

Asperger syndrome is commonly defined as a pervasive developmental disorder (PDD). PDDs are defined as a group of behavioral disorders that have two common features: problems in social interaction and problems with communication, both verbal and nonverbal.

# Description

Children with AS are not mentally retarded. They learn to talk at the usual age and often have above-average verbal skills. They have normal or above-normal intelligence and the ability to feed or dress themselves and take care of their other daily needs. The most distinctive features of AS are problems with social interaction, particularly making friends with others; difficulties with nonverbal communication (e.g., facial expressions); peculiar speech habits that include repeating words or phrases or talking in a flat tone of voice; an apparent lack of "common sense" and a fascination with odd or obscure subjects (e.g., the parts of a clock or small machine, railroad schedules, astronomical data, etc.) that may prevent the child's developing other interests.

#### Also Known As

AS, high-functioning autism (HFA), autistic psychopathy

**Cause** Unknown

#### Symptoms

Social difficulties, restricted interests

Duration Lifelong



*Craig Morris, who suffers from Asperger syndrome.* AP IMAGES.

Although AS is not a physical disorder as such, some children with the syndrome are clumsy or make awkward repetitive physical movements. They typically find team sports and skills like swimming or riding a bicycle much more challenging than most children. They may also have strange or eccentric behaviors like hand wringing, finger flapping, or swaying in place. Some children with AS are unusually sensitive to bright lights, loud sounds, and changes in temperature.

# **Demographics**

The exact frequency of AS is not known, partly because doctors in different countries disagree about its exact definition. Some think that AS is a subtype of autism while others think it is a distinct disorder. Estimates range between one child in 250 and one child in 10,000. What is known is that the disorder affects four times as many boys as girls. AS is thought to affect all racial and ethnic groups equally.

# Hans Asperger (1906-1980)

Hans Asperger was an Austrian pediatrician who is credited with the earliest description of the disorder that now carries his name. He was born on a farm near Vienna and graduated from medical school in 1931. The next year he took a position in the University Children's Hospital in Vienna. He also worked as a psychiatrist in Leipzig in eastern Germany for several years.

In 1944 Dr. Asperger published his landmark paper on what he called autistic psychopathy in a group of four boys. He described the disorder as an unusual pattern of behaviors and abilities that included, in his words, "a lack of empathy, little ability to form friendships, one-sided conversation, intense absorption in a special interest, and clumsy movements." He called his young patients "little professors" because they could talk about their favorite subjects in great detail.

Some people who knew Dr. Asperger as a child thought that he had some characteristics of AS: he was a lonely boy who had difficulty making friends, but he was good at languages and had a special interest in an Austrian dramatist named Franz Grillparzer. Asperger used to bore his elementary school classmates by reciting long speeches from Grillparzer's plays.

# **Causes and Symptoms**

The cause of AS is not known, although some think it may be related to a decreased flow of oxygen to the baby's brain during childbirth. Another theory is that AS is genetic because it appears to run in families; however, no specific gene has yet been identified with the disorder. A team of German researchers reported in January 2008 that they had excluded a specific gene that other scientists thought might be a partial cause of AS.

The symptoms of AS vary somewhat according to the child's age. Young children with AS typically have problems picking up social cues and understanding the basics of interacting with other children. The child may want friendships but find him- or herself unable to make friends.

The symptoms of AS usually become much more noticeable during the elementary school years. It is at this point that the child's physical clumsiness, difficulty making eye contact, unusual but restricted interests, and odd behaviors increase his or her difficulty making friends. Although the child learns language easily, he or she may speak in a rapid, jerky, or overly loud way. Children with AS are also extremely

literal in their use of language; they often fail to understand the symbolic or humorous uses of language, such as the saying that "it's raining cats and dogs."

Adolescence is one of the most painful periods of life for young people with Asperger's, because social interactions are more complex in this age group and require more finely tuned social skills. In addition, teenagers with AS are often naïve and may be easily manipulated or cheated by their more sophisticated peers.

Adults with AS are usually able to complete their education and join the workforce. Some become exceptional scholars; one of the

children that Hans Asperger studied in the 1940s became a noted astronomer, publishing a paper on an error in Isaac Newton's work that he had first noticed in childhood. The chief difficulties that men with AS have in adult life are courtship and marriage. They may want very much to marry and have a family but do not understand the many social interactions that are part of the dating process leading to marriage. One man with AS became a surgeon and accomplished musician, but lived with his parents until he was fifty years old. He then married a distant cousin half his age who lived in another country; the marriage lasted only a few months.

# Diagnosis

AS is usually diagnosed when the child is between four and eleven years old. There is no laboratory or imaging test that can detect AS, although the child may be given a CT scan to rule out other disorders of the nervous system or a hearing test to rule out partial deafness. Since a diagnosis of AS is based on a pattern of behaviors rather than a set of specific physical characteristics, the child is usually diagnosed by being observed by several different doctors, including a psychiatrist and neurologist, to look for recurrent patterns in the child's speech, movements, and interactions with other people. A test that is sometimes used to screen for AS involves asking the child to carry out a task that depends on the ability to predict how another person might respond to a given situation. Children with AS usually lack the ability predict someone else's thoughts and feelings.

#### Treatment

Treatment for AS depends on the specific child's range of abilities and interests. There is no single set or series of treatments that will work for every child with AS. Some children who are very clumsy benefit from physical therapy that improves their coordination and ability to participate in sports. Some children may need to work with a speech therapist in order to learn to speak in a normal tone of voice. Teachers and parents can often help a child with AS work on his or her social skills.

Medications are not used to treat AS itself, although adults with AS may be given antidepressant medications if they become depressed. In some cases psychotherapy is useful for adults with AS who are discouraged by their condition or who have developed other mental disorders.

#### Prognosis

The prognosis of AS varies depending on the person's level of intelligence, career interests, and the amount of support he or she receives from friends and family. Divorce and family breakup can be very upsetting to children with AS and complicate their educational and social development. Career counseling can help a child with AS choose an occupation that will make the most of his or her interests and preferably allow him or her to work alone rather than as part of a team. In addition, children with AS actually have an advantage in some fields, like music or mathematics, because of their ability to spend hours developing and practicing skills in their field of interest. People with AS have become successful scientists, mathematicians, architects, medical researchers, computer scientists, musicians, and engineers. Some famous people who are thought to have had AS include such writers as Jonathan Swift and Patricia Highsmith; such philosophers as Bertrand Russell and Ludwig Wittgenstein; and such musicians as Bela Bartok, Erik Satie, and Glenn Gould.

As far as is known, people with AS have the same life expectancy as others of their age, race, or sex, but they do have a higher risk of developing eating disorders, depression, schizophrenia, or Tourette syndrome. Having another mental disorder then places them at a higher risk of suicide than people in the general population.

#### Prevention

There is no known way to prevent AS because its causes are not completely understood.

#### The Future

There are no new treatments for Asperger syndrome, nor is it known whether the disorder is becoming more common, partly because of disagreement about its definition.

One major change since 2000 is the development of an autistic rights or autistic pride community. Rather than accepting the standard definition of Asperger syndrome as a disease to be cured, some adults with AS prefer to describe it as simply having a different type of brain

# WORDS TO KNOW

- **Aspie:** An informal name for a person with Asperger syndrome.
- Autism: A developmental disorder that appears by three years of age and is characterized by limited communication skills, difficulties in communicating with others, and difficulties forming relationships. Some doctors think that Asperger syndrome is a subtype of autism.

**Pervasive developmental disorder (PDD):** A diagnostic category for a group of childhood disorders characterized by problems in communication skills and social interactions. Asperger syndrome is classified as a PDD.

**Tourette syndrome:** A neurological disorder characterized by recurrent involuntary body movements and repeated words or grunts.

organization. A Web site for "aspies," as they have nicknamed themselves, called Wrong Planet was set up in 2004. As with many other disorders, the Internet is offering people with AS the opportunity to form their own communities, find support, share their personal experiences, and learn about the latest research.

#### SEE ALSO Autism; Autism spectrum disorders; Tourette syndrome

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# **m** Asthma

# Definition

Asthma is an inflammatory disorder of the airways leading to the lungs. The disease causes the airways to constrict and produce mucus that makes breathing difficult. Asthma is considered an atopic disease along with eczema and hay fever.

# Description

The symptoms of asthma vary from person to person. Some people diagnosed with asthma have occasional episodes of mild wheezing or coughing but are able to breathe normally in between such episodes. Others may have life-threatening attacks in which they turn blue for lack of oxygen, experience severe chest pain, and may even lose consciousness.

Doctors classify asthma into four categories of severity:

- Intermittent: The patient has asthma episodes less than once a week; can breathe normally between episodes; and does not need daily medication.
- Mild persistent: The patient has episodes more often than once a week but less than once a day; the episodes sometimes affect sleep and activity levels; and the patient can control symptoms by using an inhaler.
- Moderate persistent: The patient has episodes every day; the symptoms affect sleep and activity levels; and the patient needs longacting oral medications as well as an inhaler.
- Severe persistent: The patient has continuous symptoms; is frequently disturbed at night and must limit daytime activity; needs steroid medications as well as oral drugs and long-acting inhalers.

# Demographics

Asthma is one of the most common chronic (long-term) diseases around the world and is the single most common cause of hospitalization for children in the United States. According to American Academy of Allergy, Asthma and Immunology (AAAAI), 20 million people in the United States

Also Known As Hyperactive airway

# disease Cause

Allergens, infections, exposure to smoke or air pollution, exercise, cold air, stress, some medications

#### Symptoms

Wheezing, coughing, shortness of breath, tightness of the rib cage

#### Duration

Minutes to days (episodes); disorder itself may be lifelong

#### Asthma



*Teenager using an inhaler to treat her asthma.* SHUTTERSTOCK.

have asthma, with 9 million children under the age of eighteen diagnosed with the disease. Two-thirds of all cases of asthma in North America are diagnosed in children before age eighteen.

Asthma is responsible for about 500,000 hospitalizations each year in the United States and 5,000 deaths. There are about 13 million physician office visits and 1.2 million outpatient department visits due to asthma. Children with asthma miss about 13 million days of school each year and adults with asthma miss 25 million work days. The annual costs to the American economy for asthma are \$11.5 billion for direct health care and \$5 billion for lost productivity.

Risk factors for asthma include:

- A family history of asthma.
- Living in a large city.
- Exposure to secondhand smoke in the home.
- Exposure to chemicals used in farming, manufacturing, dry cleaning, hairdressing, and other occupations.
- Race. African Americans and Puerto Ricans in the United States have higher levels of asthma than members of other races, including Native Americans and Pacific Islanders.

# Jackie Joyner-Kersee

Jackie Joyner-Kersee (1962–) is a retired Olympic champion who was voted the greatest woman athlete of all time by *Sports Illustrated for Women*. Named after former First Lady Jackie Kennedy, Joyner-Kersee began her career in sports at the University of California at Los Angeles in 1980. She participated in basketball and track and field events. Between 1984 and 1996, Joyner-Kersee won three gold, one silver, and two bronze medals in four different Olympic Games.

Joyner-Kersee was diagnosed with asthma during her freshman year of college but did not take her diagnosis seriously. Like many people treated for asthma, she felt good enough from time to time to stop taking her long-term medications on a regular basis. She had several close calls that sent her to the emergency room, then she experienced a nearly fatal asthma attack in 1993. She resolved not only to take her own health more seriously but also to travel across the United States educating children diagnosed with asthma about the importance of getting treatment. In 2007 Joyner-Kersee joined sports stars Lance Armstrong, Cal Ripken Jr., Muhammad Ali, and Andre Agassi to form a charitable organization called Athletes for Hope. Joyner-Kersee herself hosted the 2008 Winning for



Jackie Joyner-Kersee competing in a heptathalon. AP IMAGES.

Life track and field event, which draws more than 1,000 female student athletes from four Midwestern states.

- Gender. Boys are twice as likely as girls to develop asthma in childhood, but by adolescence the numbers are equal. Women are more likely than men to be diagnosed with asthma as adults.
- Obesity.
- Low birth weight.

## **Causes and Symptoms**

The basic cause of asthma is the hypersensitivity of the person's airway to some trigger in the environment. When the tissues in the airway react to the trigger, they constrict (become narrower), which reduces the amount of air that can flow into, and especially out of, the lungs. The inflamed tissues also secrete mucus, which further limits the airflow in the bronchi and adds to the wheezing, coughing, and feeling of tightness in the chest that are part of an asthma attack.

Most doctors think that genetic factors are partly responsible for the sensitivity of the lung tissues in asthma patients. It is known that the disorder runs in families. At least twenty-five different genes have been associated with an increased risk of asthma in six or more separate groups of people. Researchers still do not understand, however, exactly how these genes (or others that may be discovered in the future) interact with triggers in the environment to produce the symptoms of asthma.

Specific triggers that can produce asthma attacks in patients include:

- Allergens carried in the air. These include pollen, dust, mold, pet dander, and cockroach droppings.
- Getting a cold or other upper respiratory infection.
- Physical exercise. This is called exercise-induced asthma or EIA.
- Cold dry air.
- Tobacco smoke and other air pollutants.
- Medications, particularly aspirin, other nonsteroidal anti-inflammatory drugs (NSAIDs), and beta blockers (heart drugs).
- Foods that cause allergic reactions. Common offenders are peanuts, shellfish, and eggs.
- Gastroesophageal reflux disease (GERD).
- The menstrual cycle in some women.
- Emotional stress or tension.
- Shampoo, hair gel, fabric softener, deodorant, cologne, air freshener, shaving lotion, scented tissues, and similar personal and household products.
- Chlorinated swimming pool water.

# Diagnosis

Asthma can be difficult to diagnose, particularly in children younger than five years, because the symptoms can vary considerably in frequency and severity. In some cases, the person's symptoms may be misdiagnosed as a bad cold, pneumonia, or emphysema. To narrow the diagnostic possibilities, the doctor will take a family history as well as the patient's medical history and give the patient a complete physical examination. The next step in diagnosis is to test the patient's lung function. There are several tests that can be done:

- Spirometry. A spirometer is a machine that measures the largest amount of air that a person can breathe out after taking a very deep breath.
- Peak airflow meter. This is a device that can be used at home as part of an asthma action plan to detect minor changes in lung capacity before the symptoms of an asthma attack appear. The meter can be used in the doctor's office to measure lung function. The patient may be asked to use the meter before and after taking a dose of a bronchodilator, which is a type of drug that opens up the airway. If the patient's reading on the peak airflow meter improves after taking the bronchodilator, he or she is likely to have asthma.
- Bronchial challenge test. In this type of test, a small amount of a drug that is known to trigger asthma is given to the patient. If the patient's airway narrows after taking the drug, he or she is diagnosed with asthma. The bronchial challenge test is particularly useful in identifying patients with intermittent asthma.

### Treatment

Treatment for asthma is based on the severity and frequency of the patient's symptoms and is aimed at controlling symptoms or preventing episodes of asthma. There is no cure for the disease currently. Most asthma patients need a combination of long-term control medications and quick-relief or rescue medications. The long-term drugs are taken every day over long periods of time to prevent asthma attacks. They may take several weeks to become fully effective. The long-term medications given to control asthma include inhaled corticosteroids, long-acting inhaled bronchodilators, or oral medications like theophylline.

Quick-relief or rescue medications include oral corticosteroids and short-term bronchodilators like Atrovent or albuterol. The effects of rescue medications last between four and six hours. A patient who needs these drugs very often probably needs to have his or her long-term medications adjusted.

Asthma patients who are allergic to specific foods or airborne allergens may need treatment for their allergies as well as their asthma.

# Prognosis

About half of all children diagnosed with asthma outgrow their disease by their late teens or early twenties and do not need further treatment. Patients who develop asthma as adults are more likely to have chronic health problems related to the disease.

# Prevention

People cannot prevent getting asthma, but they can reduce the frequency and severity of asthma attacks by following the recommendations of the Centers for Disease Control and Prevention (CDC):

- Draw up an asthma action plan with the family doctor. The CDC recommends the sample plan found at http://familydoctor.org/ online/famdocen/home/common/asthma/basics/696.html.
- Identify and avoid personal asthma triggers.
- Learn to identify the warning signs of an asthma attack.
- Monitor breathing periodically by taking measurements at home with a peak air flow meter.
- Treat attacks early. Quick action reduces the likelihood of a severe attack.
- Take asthma medications as directed. Do not stop taking them without consulting the doctor even though the asthma seems to be getting better.

# The Future

Asthma is a concern to public health doctors because its rates have risen rapidly in the United States since the 1960s. The number of children under the age of five diagnosed with asthma rose 160 percent from 1980 to 2006. Similar increases have been reported in other developed countries, including Canada, Australia, New Zealand, Germany, and the United Kingdom. Some of the explanations that have been offered for the increase include people moving from rural areas into cities; air pollution; increased exposure to chemicals in the environment; and increased exposure to tobacco smoke in the household. It is expected that asthma rates will continue to rise in developed countries over the next several decades.

**SEE ALSO** Eczema; Gastroesophageal reflux disease; Hay fever; Sleep apnea; Smoking

# WORDS TO KNOW

- **Allergen:** A substance that causes an allergic reaction in individuals who are sensitive to it.
- Atopic disease: Any allergic disease that affects parts of the body that are not in direct contact with the allergen. Asthma, eczema, and hay fever are all atopic diseases.
- **Bronchodilator:** A type of drug that opens up the bronchi, increasing airflow and relieving wheezing and other asthma symptoms.
- **Bronchus (plural, bronchi):** One of the two major divisions of the airway that lead into the right and left lungs.

Chronic: Long-term.

- **Peak airflow meter:** A handheld device that asthma patients can use at home to monitor their lung capacity in order to treat the warning signs of an asthma attack as soon as possible.
- **Spirometer:** A device that is used to test the air capacity of a person's lungs and the amount of air that enters and leaves the lungs during breathing.
- **Wheezing:** A continuous harsh whistling sound produced by the airways of an asthma patient when the air passages are partly blocked.

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# U

# Astigmatism

# Definition

Astigmatism is a condition in which either the cornea or the lens of the eye has an abnormal curvature, causing out-of-focus vision.

# Description

Astigmatism is a common refractive error in which a person cannot see objects with complete clarity. In some cases the person can see vertical lines more clearly than horizontal lines; this is called with-the-rule astigmatism. In other cases the person can see horizontal lines clearly while vertical lines look blurred; this is called against-the-rule astigmatism. Children are more likely to have with-the-rule astigmatism while adults are more likely to have against-the-rule astigmatism.

# Demographics

Astigmatism is a very common visual defect. It often exists together with nearsightedness (myopia) or farsightedness (hyperopia). It is estimated that as many as a third of the general American population has some degree of astigmatism. Mild astigmatism is considered normal and may not require corrective lenses.

Astigmatism is equally common in men and women and in all races and ethnic groups.

#### Cause

Inborn irregular curvature of either the cornea or the lens of the eye

#### Symptoms

Inability to form a sharply focused image of an object

#### Duration

Lifelong unless corrected by surgery



*Eye with astigmatism (top) see a distorted image, which can be changed with a corrective lens.* THE GALE GROUP.

### **Causes and Symptoms**

The exact cause of astigmatism is not known; it is usually present from birth. In a few cases, astigmatism may result from an injury to the eye or from keratoconus, an eye disorder that causes the tissue of the cornea to become thinner over time. In an astigmatic eye, the irregularly shaped cornea or lens prevents light from focusing properly on the retina, the light-sensitive tissue layer that lines the back of the eye. When the cornea is irregularly curved, the eye focuses light on two different points inside the eye rather than one. If the person is nearsighted, one of the two focal points will lie in front of the retina; if the person is farsighted, the second focal point will lie behind the retina.

In simple astigmatism, one of the two focal points will lie on the retina itself. If the person has compound myopic astigmatism, however, both focal points lie in front of the retina. Similarly, in compound hyperopic astigmatism, both focal points lie behind the retina. A few people have what is called mixed

astigmatism, in which one focal point falls in front of the retina and the other behind it.

The basic symptom of astigmatism is that objects look blurred at any distance. People who are severely astigmatic may also develop headaches, eye strain, and fatigue along with blurring of vertical, diagonal, or horizontal lines if their refractive error is not corrected.

# Diagnosis

Astigmatism and other refractive errors are evaluated by a series of vision tests. After the examiner takes a history of the patient's symptoms (including a family history of eye problems), the patient is usually asked to read the letters on an eye chart known as a Snellen chart. The examiner may also shine lights into the eyes or administer eye drops that allow him or her to see all the structures inside the eye clearly. This part of the

# Was El Greco Astigmatic?

El Greco, or "The Greek," was the nickname of Domenikos Theotokopoulos (1541–1614), a great artist of the seventeenth century. Born on the island of Crete, he studied art in Italy with Titian, another famous painter, and moved to Spain in 1577. As he grew older, the human figures in his paintings became progressively more elongated, almost looking unnatural. This lengthening is one of the most distinctive characteristics of his style, making it relatively easy for art historians to tell the difference between his paintings and those produced by other artists of the period.

Some art historians suggested that his distortion of the human figure might have been the result of astigmatism. They theorized that El Greco had a type of astigmatism that caused him to see elongated human figures as normally proportioned. Though this theory is interesting, researchers generally believe his style was purposeful, not the result of a refractive error. As evidence they state that some of the figures in his paintings have normal body proportions, and x-ray studies of his paintings reveal that he sketched normally proportioned figures on his canvas before starting to



*The painter El Greco.* © INTERFOTO PRESSEBILDAGENTUR / ALAMY.

add color and then painted his elongated figures over the sketched outlines.

examination allows the doctor to evaluate the patient for nearsightedness or farsightedness.

To determine the degree of astigmatism, the examiner will use a keratometer, a device that measures the curvature of the cornea. The examiner will measure the steepest and flattest curves on the cornea. Another technique that can be used to evaluate astigmatism is called corneal topography and uses a device called a videokeratoscope. The keratoscope part of the machine projects rings of light onto the cornea while the examiner studies the pattern of the reflected light and records it with a video camera. Corneal topography provides a more detailed picture of the shape of the patient's cornea and can be used to determine the proper fit for contact lenses.

### Treatment

Mild astigmatism may not need corrective treatment. A person should see their eye doctor, however, if they are developing headaches or eye strain, or if blurry vision is interfering with daily activities.

People whose astigmatism is severe enough to require correction have several options:

- Eyeglasses. These can be used to correct astigmatism caused by uneven curvature of the lens as well as the cornea. Eyeglasses are prescribed by an optometrist or ophthalmologist but made and fitted by an optician.
- Hard contact lenses. These usually provide more effective correction of astigmatism than soft contact lenses.
- Orthokeratology (Ortho-K). This is a procedure in which the person wears hard contact lenses for several hours overnight in order to gradually correct the curvature of the cornea. The lenses are removed during the day. Ortho-K is also referred to as corneal molding. It does not permanently improve vision; if the patient stops wearing the retainer lenses, their vision may return to its original condition.
- Laser surgery. An ophthalmologist can use lasers to reshape the cornea either by making a flap in the surface of the cornea and reshaping the tissue of the cornea under the flap, or by completely removing the upper layer of tissue in the cornea before reshaping the lower layers of tissue.

There are drawbacks to surgical correction of refractive errors, however. These include the risks of infection, development of haze in the cornea, or dry eyes. In some cases the surgeon may need to perform a second operation if the first one either overcorrected or undercorrected the shape of the patient's cornea.

It is important for a patient with astigmatism to discuss all the treatment options with the optometrist or ophthalmologist, as no two people will have exactly the same degree of visual blurring or the same lifestyle.

# Prognosis

Astigmatism may either improve over time or grow worse; it also changes from with-the-rule astigmatism in childhood to against-the-rule astigmatism in adulthood in some people. Most people with astigmatism who need corrective lenses or other types of treatment, however, do very well.

# WORDS TO KNOW

- **Against-the-rule astigmatism:** A type of astigmatism in which the eye sees horizontal lines more clearly than vertical lines.
- **Astigmatism:** A refractive error caused by irregularities in the shape of the cornea or the lens of the eye.
- **Cornea:** The transparent front part of the eye where light enters the eye.
- Hyperopia: The medical term for farsightedness.
- **Keratoconus:** An eye disorder in which the tissue of the cornea grows thinner over time.
- Myopia: The medical term for nearsightedness.
- **Ophthalmologist:** A doctor who specializes in diagnosing and treating eye disorders and can perform eye surgery.

- **Optician:** An eye care professional who fills prescriptions for eyeglasses and corrective lenses.
- **Optometrist:** An eye care professional who diagnoses refractive errors and other eye problems and prescribes corrective lenses.
- **Orthokeratology:** A treatment for astigmatism that consists of wearing hard contact lenses overnight to reshape the cornea during sleep. The lenses are removed during the day.
- **Refractive error:** A general term for vision problems caused by the eye's inability to focus light correctly.
- **Retina:** The light-sensitive layer of tissue at the back of the eyeball.
- With-the-rule astigmatism: A type of astigmatism in which the eye sees vertical lines more clearly than horizontal lines.

About 20 percent of people with keratoconus, however, will eventually need a corneal transplant.

## Prevention

Astigmatism is largely a matter of heredity and cannot be prevented. People can, however, live comfortably with it by visual screening in childhood, regular eye checkups at all ages, and wearing corrective lenses if necessary.

# The Future

Astigmatism is such a common refractive error in the general population that it is not likely to disappear in the near future. It is possible that the present variety of treatment options will be improved or expanded by further research.

SEE ALSO Hyperopia; Myopia

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Attention-Deficit Hyperactivity

Also Known As ADHD, ADD

# Cause

Unknown

#### Symptoms

Thoughtless or impulsive behaviors; difficulty paying attention and sitting still

#### Duration

Years

Disorder

U



Kids with attention-deficit hyperactivity disorder have difficulty focusing on one task if there is a distraction. © VSTOCK / ALAMY.

focusing or paying attention, are disorganized, and frequently behave in impulsive ways.

The disorder has been known to doctors for over a century. The earliest description of it was written by a German doctor in 1845, about his own son. In 1902 a British doctor named George Still gave a series of lectures to other doctors, in which he described children who would now be called hyperactive and impulsive. Still thought that the disorder might be genetic in origin. He emphasized the fact that the behavioral problems in these children did not result from bad parenting.

There are three basic types of ADHD: inattentiveness; impulsiveness; and a combination of the two.

# Description

The central characteristic of all three types of ADHD is that the child (or adult) diagnosed with the disorder has fallen behind in school and in social relationships. Children diagnosed with the inattentive type of ADHD daydream in school; do not seem to listen when someone speaks directly to them; do not follow instructions and fail to complete chores or homework assignments; have trouble with tasks that require concentration; and are in general disorganized. They are more easily overlooked by parents and teachers, however, because their behavior is not as disruptive as the hyperactivity or impulsiveness of other children diagnosed with ADHD.
Children diagnosed with the hyperactive or impulsive type of ADHD are the ones who cannot sit still in school. They call out answers or talk all the time, get up and walk around the room, or try to do several things at once. They do not think before they speak, act without considering the consequences of their actions, or have trouble taking turns with other children. Teenagers with this type of ADHD may say that they feel inwardly restless much of the time or need to find things to do to keep busy.

One important thing to keep in mind about ADHD is that the behaviors—whether inattentiveness, hyperactivity, or a combination of both—must be inappropriate to the child's age and must continue for six months or longer. Not every episode of absentmindedness or fidgeting in school means that a child has ADHD. In addition, children's personalities differ, including their level of activity and their level of interest in certain activities. For example, some children have no difficulty finishing their homework in a subject that interests them but may have trouble focusing on an assignment in a subject they do not like.

The signs of ADHD in adults are often harder to recognize than in children. Adults with ADHD are more likely to say that they have trouble relaxing than feeling that they have to race around physically. Inattentiveness may take the form of forgetting meetings, work deadlines, or social get-togethers. Impulsiveness may take the form of moodiness, temper tantrums, or impatience in traffic jams.

# Demographics

According to the National Institute of Mental Health (NIMH), between 3 and 7 percent of children in the United States have ADHD. This group would include about 2 million children as of 2008. Worldwide, the rate of ADHD in children is thought to be somewhere between 8 and 12 percent.

Boys are three to five times more likely than girls to be diagnosed with ADHD. The inattentiveness subtype is more common in girls than in boys, however. As far as is known, ADHD is equally common in all racial and ethnic groups in the United States.

In adults, the gender ratio is virtually equal. The rate of ADHD in American adults is thought to be between 2 and 7 percent.

# **Causes and Symptoms**

The causes of ADHD are not completely understood, but studies of brain function using positron emission tomography (PET) scans indicate that the brains of children with ADHD may be structured differently from those of children without the disorder. More specifically, some researchers think that the parts of the brain that govern attentiveness do not have normal levels of dopamine, a chemical produced by the brain that allows nerve cells to transmit signals from one cell to another. The fact that medications that stimulate the release of dopamine are beneficial to children with ADHD supports this theory.

There are also genetic factors involved in ADHD even though no specific gene has been identified as a cause of the disorder. It is known that the disorder runs in families. The parents and siblings of children with ADHD are two to eight times more likely to develop ADHD than the general population.

Some researchers think that environmental toxins, particularly lead, and a mother's drinking or smoking during pregnancy are risk factors for ADHD in the child. There is also some evidence that very low birth weight or premature birth may be risk factors for ADHD. It is not known whether the emotional climate in the child's family contributes to the disorder, although some doctors think that mood or anxiety disorders in other family members may make the child's symptoms worse. Other unproven theories include the idea that ADHD is caused by head injuries, allergic reactions to food additives, or a diet high in sugary foods.

Symptoms of inattentiveness related to ADHD include:

- Failing to pay close attention to details; frequently making careless mistakes.
- Having difficulty paying attention to tasks for more than a few minutes.
- Appearing not to listen when spoken to.
- Not following through on instructions and failing to complete homework or other tasks.
- · Having difficulty organizing tasks.
- Frequently losing pencils, books, or other items necessary to complete homework or other tasks.
- Being forgetful.
- Being easily distracted.

Symptoms of impulsiveness and hyperactivity include:

- Fidgets or squirms in seat.
- Leaves classroom seat and runs around.

- Has difficulty playing quietly.
- Seems to be constantly on the go.
- Talks a lot; finds it hard to keep quiet.
- Blurts out answers in class.
- Finds it hard to take turns or wait in line.
- Butts in on conversations or intrudes on the privacy of others.

# Diagnosis

The diagnosis of ADHD is complicated, not only because the child's age, overall personality, and medical history must be taken into account, but also because about 45 percent of children with ADHD have at least one other psychiatric disorder, most commonly anxiety disorders, depression, Tourette syndrome, or bipolar disorder. There is no single laboratory test, imaging study, or neurological test that can be used to diagnose ADHD, although there are behavioral checklists and questionnaires that doctors can use to narrow the diagnostic possibilities. The diagnosis usually follows a series of interviews with psychiatrists and psychologists as well as the child's pediatrician. A complete physical examination is important to rule out vision problems, hearing loss, seizure disorder, or hyperthyroidism as possible causes of the child's behavior.

The diagnosis of ADHD in children is based on whether the child meets the criteria specified in a diagnostic manual on mental disorders:

- The symptoms must have started before the child was seven years old.
- The child must have had the symptoms over a period of at least six months.
- The child must have six or more of the signs or symptoms of inattentiveness or hyperactivity/impulsiveness listed in the previous section.
- The symptoms must be shown to have affected the child's ability to function in at least two areas of life, usually home, relationships with friends, and schoolwork. Thus, a child who is impulsive or inattentive at school but relates well to friends or other family members would not be diagnosed with ADHD.

A set of criteria called the Wender Utah Rating Scale is commonly used to diagnose ADHD in adults:

- A history of ADHD in childhood
- · Hyperactivity and poor ability to focus or concentrate

- Difficulty completing tasks
- Mood swings
- Difficulty controlling temper
- Inability to handle stress
- Impulsive behaviors

#### Treatment

Treatment for ADHD usually includes a combination of medications and behavior therapy. The use of medications has been controversial. However, children treated with both medications and behavior therapy have been found to do better than those treated with behavior therapy alone. It is important to understand that the medications prescribed for ADHD do not cure the disorder. The medications only help the child control the symptoms, and they must be taken every day.

The medications most commonly used to treat ADHD are psychostimulants, which work by targeting the parts of the brain that produce dopamine. These drugs include Ritalin, Adderall, and Dexedrine. More recently, the Food and Drug Administration (FDA) approved the use of a non-stimulant medication called Strattera, which works on another brain chemical called norepinephrine. Norepinephrine is a hormone that affects a person's ability to pay attention and respond appropriately to stress. Although medications for ADHD generally work well, they do have side effects, such as sleep problems, weight loss, appetite loss, and nervousness.

In addition to behavior therapy, children with ADHD may receive social skills training and attend support groups. Many doctors recommend parenting skills training for the parents of children with ADHD and family therapy for the entire family.

Adults with ADHD are also treated with a combination of drugs and psychotherapy. However, because adults need larger doses of psychostimulants than children do, they must see their doctor periodically to make sure that the drugs are not interfering with other medications they may be taking, particularly drugs to control high blood pressure.

Some forms of alternative medicine that have been suggested as treatments for ADHD include special diets, vitamin supplements, and various herbal preparations. There is no evidence, however, that any of these alternative approaches are useful for children or adults with the disorder.

# WORDS TO KNOW

**Dopamine:** A brain chemical that helps to regulate movement.

**Norepinephrine:** Another brain chemical that affects a person's ability to pay attention.

**Psychostimulant:** A type of drug that increases the activity of the parts of the brain that produce dopamine.

#### Prognosis

Doctors think that between 30 and 70 percent of children diagnosed with ADHD will continue to have some symptoms of the disorder as adults. Most children diagnosed with the disorder benefit from appropriate treatment, although they have a higher risk of alcohol and drug abuse in adolescence. As many as 65 percent may continue to have problematic symptoms of ADHD that keep them from reaching their full academic or work potential in adult life.

#### Prevention

There is no known way to prevent ADHD because the causes of the disorder are still not completely understood.

## The Future

Doctors are not certain whether the rate of ADHD in the American population is likely to increase or remain at its present level. Current research on the disorder includes studies comparing newer drugs for ADHD to older psychostimulants; studies of ADHD in adults; studies of the relationship of childhood ADHD to substance abuse in adolescence and adulthood; and evaluations of new diagnostic questionnaires for ADHD.

**SEE ALSO** Bipolar disorder; Lead poisoning; Prematurity; Seizure disorder; Tourette syndrome

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# **U** Autism

# Definition

Autism is classified by the American Psychiatric Association as a pervasive developmental disorder (PDD). It is characterized by difficulties in social interaction; problems with verbal and nonverbal communication; and

#### Autism



Boy at a boarding facility for severly autism children, in Montmoreau, France. PHANIE / PHOTO RESEARCHERS, INC.

#### Also Known As

Pervasive developmental disorder, PDD

# Cause

Unknown

#### Symptoms

Poor social development and communication, restricted interests, repetitive behaviors

**Duration** Lifelong patterns of repetitive behaviors coupled with a narrow range of interests—all beginning before the child is three years old. Autism is considered a spectrum disorder, because children diagnosed with it vary widely in the severity of their symptoms; some may be only mildly affected and eventually live independently, while others suffer from severe mental retardation and require lifelong care.

# Description

Though defined as a disorder for less than a century, autism has certainly existed for a much longer period of time. The term *autism* itself appears to have been coined independently by Hans Asperger (1906–1980), an Austrian pediatrician who first used it in a German-language medical journal in 1938, and Leo Kanner (1894–1981), an Austrian psychiatrist who had moved to the United States and published a paper on what he called infantile autism in 1943. Since the 1940s, the definition has expanded to include children who might

have simply been described as odd or strange in the 1960s, as well as children who are severely disabled by the disorder.

With regard to social interaction, children with autism are commonly described as loners who are unaware of other people's feelings. They may not respond to their names or make eye contact, and they dislike being held or cuddled. In terms of behaviors, autistic children prefer sameness and are upset by change. They may make repetitive gestures like flapping their hands, spinning, or rocking back and forth. They are often unusually sensitive to lights, noises, or touch, but at the same time they may be completely unaware of danger. Many autistic children seem to be insensitive to pain and do not tell their parents when they are hurt or sick.

Language skills are the third major area in which autistic children are different from others. They often start talking later than other children and may speak in a mechanical, sing-song, or robot-like voice. A child with autism may repeat someone else's words or sentences but may not know how to form a sentence of their own. He or she may have trouble asking for something in words, using gestures or pointing instead. In some cases children with autism do not refer to themselves as "I" or "me," but use their names instead.

# Demographics

There has been considerable controversy in recent years because the rate of autism in the United States appears to be rising fairly rapidly. For many years, doctors estimated that autism affected only four or five children in every 10,000. When the Centers for Disease Control and Prevention (CDC) released a report in February 2007 stating that the rate of autism among American children had increased to one in 150 (and one in every 94 boys), many people were quite upset. Although the CDC acknowledges that some of the increase is due to changes in the definition and classification of autistic spectrum disorders or ASDs, some is likely to represent a true increase in the number of affected children. Autism is presently the second most common developmental disability in the United States after mental retardation. The CDC estimates that there are about 560,000 persons in the United States between the ages of one and twenty-one years who have an ASD.

Boys are more likely than girls to have autism; the male/female ratio in the United States is four to one. The reason for the gender difference is not known. Autism is thought to be equally common in all racial and ethnic groups in the United States.

# **Red Flags for Autism**

Although not every child who might be autistic will have all the following characteristics, the American Academy of Pediatrics (AAP) drew up a list in 2004 of "red flags" for parents to watch for:

- The child does not speak as well as others of the same age.
- The child's eye contact is poor.
- The child seems to be in his or her own world.
- The child ignores or "tunes out" other people.
- The child does not smile in return when smiled at.
- The child seems unable to communicate what he or she wants, and either leads the parent to the object or gets it him- or herself.
- The child has trouble following simple directions.
- The child does not show the parent things that he or she finds interesting.
- The child has unusually long and severe temper tantrums.
- The child prefers to play alone.
- The child (if older than two) does not pretend or play make-believe.
- The child is unusually attached to mechanical gadgets or hard objects like key chains rather than to cuddly items like blankets or stuffed toys.

The Autism Society of America estimates that the lifetime cost of caring for an autistic person ranges from \$3.5 to \$5 million, and that autism costs the United States an average of \$90 billion each year (including special education, housing, and transportation services as well as direct health care costs).

# **Causes and Symptoms**

The causes of autism are not well understood. In the 1950s many psychiatrists thought that autism was caused by cold or distant parenting— "refrigerator mothers" was a common term—and blamed parents for the disorder. This theory has been disproved by careful studies of families with autistic children. In the 1990s some people theorized that autism is caused by childhood immunizations, particularly the measles-mumps-rubella or MMR vaccine. In 2007, however, a report published in the *New England Journal of Medicine* showed that vaccines do not cause autism; it is simply that the early symptoms of autism often appear at the age when children are given the MMR vaccine.

Other possible causes of autism that are still being investigated include:

- Genetic factors. It is known that parents of an autistic child have an increased risk (one chance in twenty) of having a second child with the disorder. No specific gene or genes associated with autism has been identified, however.
- Viruses or other disease agents. Women who have rubella during pregnancy have an increased risk of having an autistic child.
- Father's age. One study showed that fathers over forty when their child was born are six times as likely to have an autistic child as fathers who were thirty or younger. The mother's age doesn't seem to make a difference.
- Difficult pregnancy or childbirth. Some doctors think that damage to a baby's brain before or during birth may play a role in autism.
- Exposure to pesticides and other toxic chemicals. There is some evidence from two studies done in California in 2007 that women exposed to certain pesticides during the first three months of pregnancy have an increased risk of having autistic children.

Common symptoms of autism have already been described; others are listed in the sidebar. Conditions that are often found in children with autism include:

- Mental retardation. About 75 percent of children with autism are mentally retarded, which complicates diagnosis of the disorder.
- Seizures.
- Self-injury or aggressive behaviors toward others.

• A loss of previously attained language or social skills. The child appears to develop normally until about fourteen months of age and then starts losing his or her ability to talk and interact with others.

# Diagnosis

The diagnosis of autism is complex; there is no single diagnostic test for the disorder. In many cases the parents start noticing problems in the child's speech and interactions with them when the child is between fourteen and eighteen months old. The diagnosis may not be made, however, until the child is two to three years of age because of the number of evaluations that may be needed. It usually takes a team of experts in child development to make the diagnosis, including a psychiatrist, a neurologist, a speech therapist, and a pediatrician, all of whom should have specific experience in evaluating and treating children with autism. There are several checklists and screeners for symptoms of autism that these health care professionals may use. In addition, they may order imaging studies, a hearing test, or (in a few cases) a blood test to rule out other possible causes of the child's behaviors.

Even though the diagnosis of autism can be complicated, it is important for parents to have their child evaluated as soon as they notice indications that the child may be autistic. The reason for early evaluation is that it improves the child's chances of making good progress in therapy. Studies have shown that children who begin treatment at or before age three do better than those diagnosed and treated later.

## Treatment

There is no cure for autism. Treatment usually involves several different types of therapy. An important form of therapy is behavior and communication therapy, which is aimed at improving the child's ability to control problem behaviors and to interact with others. In some cases the parents are taught new ways of interacting with their child while he or she is still a toddler. After the child enters school, highly structured education programs have been shown to increase the child's social as well as communication skills.

Medications may be prescribed to treat seizures, depression, anxiety, and other behavioral problems that the child may have.

# WORDS TO KNOW

**Neurologist:** A doctor who specializes in diagnosing and treating disorders of the nervous system.

**Spectrum disorder:** A disorder whose symptoms vary in severity from one patient to the next.

Because autism is a disorder that places heavy emotional as well as financial burdens on other members of the family, many families with autistic children benefit from family therapy or joining a support group.

# Prognosis

The prognosis of autism depends largely on the child's intelligence level and symptom severity. Some autistic children with above-average intelligence and relatively mild symptoms have successfully completed college, married, and started families; severely retarded autistic children, however, may require lifelong care at home or in an institution. Autistic children who start losing previously acquired language skills before age three are at increased risk of seizures as they grow older.

# Prevention

There is no known way to prevent autism.

# The Future

Researchers are looking in a number of different directions for new treatments for autism and improved diagnostic techniques. Some are studying serotonin, a chemical in the brain that affects the transmission of impulses from one cell to another, to see whether the high levels of this chemical in the blood of some children with autism are related to the disorder in some way. Other scientists are continuing to look for genes that may be linked to autism. In terms of behavior, researchers are testing the effectiveness of a program that combines parent training and medication to reduce the aggressive or disruptive behavior of some children with autism.

**SEE ALSO** Asperger syndrome; Rubella; Seizure disorder

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# U

# Autism Spectrum Disorders

Autism spectrum disorders, or ASD, is a term used by psychiatrists to refer to a group of developmental disorders of childhood that are still not well understood. The three major forms of ASD are Asperger syndrome, autism, and pervasive developmental disorder not otherwise specified, or PDD-NOS. These three disorders are sometimes grouped together with two other very rare disorders as pervasive developmental disorders or PDDs.

What the three ASDs have in common is impaired ability to communicate and interact socially with other people, combined with a limited set of interests. Children with Asperger syndrome differ from those diagnosed with autism or PDD-NOS in that they do not show a delay in language development.

The autistic spectrum disorders are thought to affect six to seven of every 1,000 school-age children. Most of these children are diagnosed as having PDD-NOS; autism accounts for one or two children in every 1,000, and Asperger syndrome for one child in every 5,000–10,000.

Most children with ASDs benefit from individualized special education programs and behavioral therapy to help them learn social skills. Many—perhaps as many as 10 percent—of these children have special gifts or talents that enable them to excel in fields that do not require working closely with others.

SEE ALSO Asperger syndrome; Autism



# Avian Influenza

# Definition

Avian influenza, or bird flu, is a form of flu caused by a strain of the influenza A virus that ordinarily affects only birds. Avian influenza is considered a zoonosis because it can be transmitted from animals to humans. It is also considered an emerging disease because it has been identified as a major public health problem within the last twenty years and threatens to get worse in the near future.

Wild birds carry many types of the influenza A virus in their digestive tract but do not usually get sick from it. Infected wild birds can, however, infect chickens, turkeys, and other domesticated birds through contact with their nasal secretions or droppings. The infection can be spread by contaminated bird feed, water, or soil as well as by direct contact between

#### Also Known As

Bird flu, H5N1 infection, influenza virus A infection

#### Cause

Influenza A virus

#### Symptoms

Cough, fever, sore throat, muscle aches, conjunctivitis

**Duration** One to two weeks wild birds and domesticated poultry. There are two subtypes of avian flu in birds: a relatively mild form that causes symptoms such as feather loss and a severe form that can kill entire flocks of chickens, geese, or turkeys within forty-eight hours.

Most cases of avian flu in humans as of 2008 have occurred in farmers or other people who were exposed to sick birds. There are very few cases of human-to-human transmission of this form of influenza. The primary reason for concern is that one specific form of the flu virus that causes avian flu—H5N1—could cause widespread sickness and death if it mutates into a form that can spread easily and directly from human to human. It was a different form of the influenza A virus (H1N1) that caused the flu pandemic of 1918–1920, which took the lives of millions of people worldwide.



# Description

Avian influenza is similar to seasonal influenza. The person develops a cough, a fever, an inflammation of the tissues lining the eyelid, a runny nose, and a headache. The incubation period in humans varies between two and seventeen days, with five days being the most common length of time. In fatal cases, the patient begins to have trouble breathing and may cough up blood about five days after symptoms of illness appear. Death usually results from viral pneumonia.

Some patients also have abdominal pain, vomiting, and bloody diarrhea in addition to respiratory symptoms.

# **Demographics**

The first cases of bird flu in humans were reported in Hong Kong in 1997. Of the eighteen people taken ill, six died from the infection. The disease has spread westward among the bird populations of Asia, Europe, and northern Africa, with cases of human illness following. In Europe, however, there have been instances of infected birds discovered without any cases of human disease. As of September 10, 2008, the World Health *Magnified image of the avian influenza virus.* © PHOTOTAKE INC. / ALAMY. Organization (WHO) reported that 387 confirmed cases of avian influenza in humans have occurred since 2003; more than half of the patients died.

There have been no confirmed cases of avian influenza caused by the H5N1 subtype in the United States in either birds or humans as of September 2008. There were, however, outbreaks of a different flu virus among American poultry in 2003 and 2004. Given the fact that the broadened spread of any subtype of flu virus increases the danger to public health in all countries around the world, many researchers think that it is only a matter of time until the H5N1 type of avian flu reaches the United States.

The risk factors for getting bird flu are not completely understood, although contact with infected birds or their droppings appears to be the single most important factor.

- People of all races and both genders appear to be equally likely to be infected.
- According to WHO, people between the ages of ten and thirtynine are most likely to die from avian flu if they become infected.
- As of 2008, 50 percent of reported cases have been in people twenty years of age or younger; 40 percent of cases have been reported in people between twenty and forty years of age.

# **Causes and Symptoms**

The cause of avian influenza in humans is a variant of the H5N1 influenza virus that has somewhat adapted itself to infecting people. There is some evidence that the same strain that infects birds also infects pigs and can jump the species barrier into humans from pigs as well as birds.

Avian influenza may affect the lower airway in humans more severely than seasonal influenza. The most common symptoms are:

- Fever above 100.4°F (38°C)
- Cough
- Sore throat
- Difficulty breathing
- Headache
- Conjunctivitis (inflammation of the tissues lining the eyelid)

#### Diagnosis

Diagnosis of suspected cases of avian flu was complicated until 2006 because samples of the patient's throat secretions had to be sent to state

health laboratories or the Centers for Disease Control and Prevention (CDC) in Atlanta, Georgia, for analysis. In February 2006, however, the Food and Drug Administration (FDA) approved a new test for diagnosing strains of bird flu in people who might have the virus. The test is called the Influenza A/H5 (Asian lineage) Virus Real-time RT-PCR Primer and Probe Set. The test gives results within four hours.

# Treatment

The CDC recommends two antiviral drugs, Tamiflu and Relenza, for treating any suspected case of bird flu in the United States. It is not known, however, whether the H5N1 influenza virus might become resistant to these drugs. In addition, the drugs must be taken within two days of the appearance of symptoms to be fully effective. Two experimental antiviral drugs were being tested in Russia and China in 2008, but neither had yet become available in the United States.

Patients with a severe case of avian influenza may need to be placed on a respirator. In addition, public health officials currently recommend that patients suspected of having avian flu should be placed in isolation to reduce the risk of the infection spreading to other people.

# Prognosis

Avian influenza in humans is a serious disease, with a mortality rate close to 60 percent. There is little information available about the long-term effects of the illness on survivors.

#### Prevention

There was no effective vaccine against avian influenza that was available to the general public in late 2008, although some were undergoing clinical trials. One of the difficulties of producing a vaccine against this type of flu in large quantities is that the virus is so powerful that it cannot easily be cultured in chicken eggs, which is the usual method for making vaccines. In June 2008, the *New England Journal of Medicine* reported on an experimental vaccine that produced a satisfactory immune response in 75 percent of the subjects who were injected with it. The vaccine appeared to be safe for widespread use in humans and could be made in cell cultures rather than in eggs.

Until an effective vaccine for avian flu is available, the most important preventive measures people can take are to wash their hands

# WORDS TO KNOW

Avian: Pertaining to birds.

**Emerging infectious disease (EID):** A disease that has become more widespread around the world in the last twenty years and is expected to become more common in the future. **Pandemic:** A disease epidemic that spreads over a wide geographical area and affects a large proportion of the population.

Zoonosis (plural, zoonoses): A disease that animals can transmit to humans.

frequently and avoid open-air markets, raw or undercooked eggs or poultry, and close contact with live poultry when visiting Asia or other countries where cases of avian flu have been reported. Travelers may also want to ask their doctor about getting a flu shot before they take their trip. Standard flu shots will not prevent bird flu, but they can protect people against being infected with seasonal flu.

## The Future

Because of the potential disaster of an avian influenza pandemic, researchers are working on developing new antiviral medications and effective vaccines as quickly as possible. No one knows if or when the H5N1 virus will mutate into a form that can be easily transmitted from one person to another. Public health officials around the world are hoping that they will be prepared if that mutation does occur.

**SEE ALSO** Influenza; Pneumonia; Severe acute respiratory syndrome

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# B



#### Also Known As

Bipolar affective disorder, manic-depressive disorder

#### Cause

Unknown; possibly genetic

#### Symptoms

Extreme shifts in a person's mood, energy level, and ability to function

#### Duration

Lifelong after adolescence

	-	

# **Bipolar Disorder**

## Definition

Bipolar disorder is the name of a group of mood disorders characterized by alternation between periods of high energy, known as mania, and periods of depression. These emotional highs and lows are much more extreme than the mood changes that most people experience. There are three major subtypes of bipolar disorder:

- Bipolar disorder I (BPI): The person has had at least one manic episode, with or without an episode of depression.
- Bipolar disorder II (BPII): The person has had at least one episode of depression and at least one hypomanic episode. Hypomania is a milder form of mania that does not interfere with the person's daily functioning.
- Cyclothymia: Cyclothymia is a mild form of bipolar disorder in which the person's highs and lows are not as extreme as in the first two types.

# Description

People with bipolar disorder alternate between periods of high energy or irritability in which they may have difficulty sleeping, act impulsively, and make ambitious plans alternating with periods of depression in which they may feel guilty and hopeless, feel unable to accomplish anything,

#### **Bipolar Disorder**

The scopolamine patch, which delivers medicine usually used for motion sickness, is now an experimental treatment for bipolar disorder. AP IMAGES.



and consider suicide. Some patients have a mixed state, in which the high energy of the manic phase of the disorder is combined with the blue mood of the depressed phase. The mixed state is more common in children or adolescents with the disorder than in adults. A person who has four or more episodes of alternating between manic and depressive phases within a twelve-month period is said to have rapid cycling bipolar disorder. Rapid cycling is more likely to develop later in the disorder. Some people with rapid cycling have several episodes of mood changes within a week or even within a single day.

Bipolar disorder takes a heavy toll on a young person's educational development, employment, and relationships. In fact, it is often misdiagnosed because people tend to attribute the person's depression and other mood changes to failed relationships or trouble in school rather than seeing the mood disturbances as the cause of these problems. In some cases, the person with bipolar disorder may be misdiagnosed as having a drug or alcohol problem.

Most people with bipolar disorder are able to function normally between episodes once they get treatment. A small minority, however, have chronic symptoms that do not improve in spite of treatment.

# Demographics

The National Institute of Mental Health (NIMH) estimates that about two million Americans over the age of eighteen have bipolar disorder and

# Bipolar disorder and suicide: one survivor's story

People with bipolar disorder have a 25-50 percent risk of attempting suicide at some point in adult life. Many of these attempts are spur of the moment rather than planned. One woman in San Francisco told a newspaper reporter of her impulsive decision to jump off the Golden Gate Bridge one afternoon in 1978. She left her apartment after lunch and drove to the bridge. "It was overcast, windy and kind of scary," she recalled later. "I was up there a long time." She was looking for the best place to jump when two police officers spotted her and asked what she was doing. When she admitted that she was going to jump from the bridge, they drove her to a hospital. "It was totally impulsive," she said years later. "I had left my cat behind, and I loved my cat."

She is grateful to the officers for saving her life that day. Diagnosed with bipolar disorder, she is careful to take her medications and to keep her depressive moods under control. "I still feel suicidal on a regular basis, but now I also have cognitive tools to deal with it," she said. "I have learned how to not act on the [suicidal] urges."

BPII. Most people develop symptoms of the disorder in the late teen years or early twenties, but some begin to show signs of the disorder in childhood and others may develop symptoms in their later years. In general, however, a person who has their first manic episode after age fifty should be examined for a medical disorder first before being diagnosed with bipolar disorder.

that the disorder costs the country about \$15.5 billion every year. About 0.8 percent of the general adult population has BPI and 0.5 percent has

Men and women are at equal risk of developing BPI, although women are more likely than men to have rapid cycling. Women are at greater risk than men of developing BPII. Bipolar disorder occurs with equal frequency in all races and ethnic groups, as far as is known.

#### **Causes and Symptoms**

The causes of bipolar disorder are not completely understood, although the disorder is known to run in families. The disorder is not caused by one gene, however, because identical twins of patients diagnosed with the disorder do not always develop it.

Some doctors think that patients with bipolar disorder may have chemical imbalances in the brain that affect moods and emotions, while others think that there may be structural differences in these patients' brains as well. Still other researchers think that bipolar disorder may be triggered by a combination of genetic factors and life experiences, as episodes of mania in some patients are known to be triggered by changes in medications, by thyroid disorders, or by inadequate sleep.

The symptoms of the manic phase of bipolar disorder may include:

- Unusually high levels of energy
- Euphoria (exaggerated sense of well-being), unrealistically high self-esteem, poor judgment
- Rapid speech, racing thoughts, insomnia
- Risky or aggressive behavior, spending sprees, increased sexual drive, drug or alcohol abuse
- Easily distracted, unable to concentrate
- Generally jumpy or agitated

The symptoms of the depressive phase may include:

- Thoughts of suicide
- Feelings of sadness or hopelessness
- Anxiety and guilt
- Loss of appetite
- · Loss of interest in friends or normally pleasurable activities
- Irritability
- Chronic pain without an obvious physical cause

Some patients with bipolar disorder have psychotic episodes, which means that they have hallucinations and other signs of losing contact with reality. These patients are frequently misdiagnosed as having schizophrenia, another severe mental illness.

# Diagnosis

There is no way to diagnose bipolar disorder through a blood test or through imaging studies of the brain. The diagnosis is made on the basis of the patient's symptoms and their history, including a family history. The doctor will give the patient a complete physical examination to rule out such physical disorders as diabetes or anemia, and a blood test to rule out thyroid disease. The patient will also be given several psychological tests to help the doctor evaluate their feelings and behaviors. Family members and friends may also be asked about the patient's recent symptoms and behavior.

# Treatment

Treatment of bipolar disorder is a complicated and lifelong process. It is important for patients to have regular appointments with a psychiatrist, a doctor who is licensed to prescribe medications for mental disorders as well as provide psychotherapy. Patients must see the psychiatrist even when they are feeling better between episodes in order to prevent relapses. In addition, psychiatrists are knowledgeable about the many different drugs that can be used to treat bipolar disorder and can replace a drug that is not working well for a particular patient with one that may be more helpful or has fewer side effects. Sometimes patients may have one set of drugs to take during a manic episode and a different set to take during the depressive phase of the illness. The groups of drugs most commonly prescribed for patients with bipolar disorder are mood stabilizers, antidepressants, anti-seizure drugs, tranquilizers, and drugs to treat psychotic episodes.

In addition to medications, patients with bipolar disorder usually work with a social worker or a psychotherapist because the disorder affects so many areas of life. Learning about the illness and how to cope with it is a crucial part of treatment; patients may be taught stress management techniques or relaxation techniques in order to help them cope more effectively with mood swings.

In some cases family therapy may be recommended so that the patient's family members can better understand the illness and not blame themselves for causing it. In addition, families are often angry at the patient because they may have to deal with the consequences of the patient's behavior, such as wild spending sprees or arrests for drug abuse, and the family members may need help in managing their anger. Patients with bipolar disorder may be hospitalized for treatment if they are judged to be a danger to themselves or others. They can be treated with electroconvulsive therapy (ECT) in the hospital if they are having a severe episode of depression. Most treatments for bipolar disorder can be given on an outpatient basis, however.

# Prognosis

The prognosis for patients with bipolar disorder is generally good as long as they keep in regular contact with their psychiatrist and follow all treatment recommendations. Men, however, appear to have a somewhat worse prognosis than women, as do patients with a history of alcohol and drug abuse.

Patients with either BPI or BPII should report all changes in mood to their doctor at once so that their treatment plan can be adjusted. Keeping a daily chart of moods and feelings that can be shared with the doctor is often helpful. Most patients are able to maintain a good quality of life in spite of the disorder; however, about 11 percent will eventually succeed in committing suicide.

## Prevention

There is no known way to prevent bipolar disorder, because the causes of it are not yet fully understood.

#### The Future

The NIMH is presently conducting several clinical trials of new medications and treatment strategies for bipolar disorder. Some of these are known as "real-world" clinical trials, because they enroll patients who are not hospitalized and are living productive lives. The newest large-scale study sponsored by the NIMH is called the Systematic Treatment Enhancement Program for Bipolar Disorder, or STEP-BD.

**SEE ALSO** Depression; Stress

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# WORDS TO KNOW

Affective disorder: A type of mental disorder characterized by disturbed emotions and feelings rather than problems with memory, thinking, or learning.

Cyclothymia: A mild form of bipolar disorder.

- **Electroconvulsive therapy (ECT):** A form of treatment for severe depression that consists of passing a low dose of electric current through the patient's brain under anesthesia.
- Euphoria: An exaggerated feeling of well-being.
- **Hypomania:** A less severe form of mania that does not interfere with normal functioning.

Mania: The high-energy phase of bipolar disorder.

**Mixed state:** A condition in which a person with bipolar disorder has the energy of the manic phase of the disorder combined with the hopeless and sad mood of the depressed phase.

**Psychosis:** Severe mental illness marked by hallucinations and loss of contact with the real world.

**Rapid cycling:** Four or more episodes of illness within a 12-month period.

**Relapse:** Recurrence of an illness after a period of improvement.

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# Bird Flu

See Avian influenza.



## Definition

Brain tumors are abnormal growths of tissue within the brain; about 150 different types have been identified. Brain tumors can be primary or secondary. Primary tumors are tumors that start to grow within the brain. Some are benign, meaning that they are not cancerous. Other primary tumors are malignant, or cancerous. Primary brain tumors are less common than secondary tumors, with most occurring in adults rather than children.

Secondary or metastatic tumors are cancers that start in other organs—most often the breasts, colon, kidneys, lungs, or skin—and then spread to the brain. Most brain tumors are metastatic cancers rather than primary tumors; about a quarter of all adults being treated for cancer in the United States eventually develop metastatic brain tumors. It is rare, however, for primary tumors of the brain to spread to other parts of the body.

#### Description

Brain tumors vary considerably in their location in the brain, their speed of growth, whether they are benign or malignant, and the specific symptoms they produce. Brain tumors can cause damage by direct destruction of normal brain tissue, by producing inflammation, or by growing so large that they increase pressure inside the skull and interfere with the functioning of other parts of the brain. Primary brain tumors can develop within the brain tissue itself, within the meninges (the membranes that cover and protect the brain and spinal cord), or within the nerves that supply the brain and spinal cord.

In adults, the most common types of brain tumors are gliomas and meningiomas. Gliomas, which account for 78 percent of malignant brain tumors in adults, arise from glial cells, which are the supportive cells in brain tissue that nourish nerve cells and hold them in place. Meningiomas are tumors that arise in the meninges. Ninety percent of mengiomas are benign; however, these tumors can still cause severe complications and death because of their location.

The most widely used system for grading brain tumors was introduced by the World Health Organization (WHO) in 1993. Grading

#### Also Known As

Brain cancers; gliomas, meningiomas, and other specific names

#### Cause

Mutations in normal brain cells; exposure to radiation; genetic diseases

#### Symptoms

Headache, nausea or vomiting, vision and speech problems, confusion, seizures, personality changes

#### Duration

Months to years

#### **Brain Tumors**



MRI scan of a brain showing a tumor in yellow. © SIMON FRASER/SCIENCE PHOTO LIBRARY, PHOTO RESEARCHERS, INC.

is based on the tumor's size, its location in the brain, and how fast it grows.

- Grade I. These are slow-growing tumors that may not produce symptoms for many years. They do not usually spread to other parts of the brain and can often be removed completely by surgery.
- Grade II. These tumors are also relatively slow-growing. They can, however, start to grow more rapidly and may spread into other parts of the brain.

- Grade III. Tumors in this category are always malignant and spread rapidly into surrounding tissue. Their cells look different from those in healthy surrounding tissue.
- Grade IV. These tumors invade other tissues very quickly and are difficult to treat. Like Grade III tumors, their cells look very different from healthy brain cells.

# Demographics

About 360,000 persons in the United States are living with a primary brain tumor as of 2008, according to the National Institutes of Health, with about 44,000 new cases of primary tumors reported each year. Primary brain tumors account for about 2.5 percent of cancer deaths in the United States each year—about 13,000 adults and children. About 100,000 Americans die each year of metastatic brain tumors.

Brain tumors are the most common type of solid tumor in children, and the second most common type of cancer in those below age fifteen. Between 15 and 25 percent of all cancers in children are brain tumors. The most common type of brain tumor in children is called a medullablastoma, a highly malignant cancer that develops in the cerebellum (the part of the brain just above the brain stem). This type of tumor is rare in adults.

Most types of brain tumors are equally common in men and women; however, meningiomas occur slightly more often in women than in men. Medulloblastomas are more common in boys than in girls. And although most brain tumors are more common in Caucasians, meningiomas are more common in African Americans than in members of other races.

Risk factors for brain tumors include:

- Age. Most brain tumors, whether benign or malignant, occur in adults over age forty-five. Brain tumors in children are rare within the first year of life; they occur most often in children between five and eight years of age.
- Exposure to nuclear radiation or radiation therapy for another type of cancer.
- Family history of brain tumors.
- Exposure to certain chemicals.

# **Causes and Symptoms**

The causes of primary tumors of the brain were not completely understood as of 2008; most researchers, however, think that defective genes are responsible. Other theories include viruses, exposure to certain chemicals, and disorders of the immune system.

Secondary brain tumors result from the spread of cancer that began elsewhere in the body, with lung cancer the most common type. Brain tumors can also develop from cancerous cells from the breast, kidneys, colon, or skin. The cancerous cells break off from the primary tumor and travel to the brain through the bloodstream.

The symptoms of a brain tumor may result from a primary tumor; they may also be the earliest symptoms of a cancer that developed in another organ. The most common warning signs include:

- Headaches, which may be most severe in the morning, new for the person, or grow worse over time
- Nausea and vomiting
- Problems with eyesight, such as seeing double or having blurred vision
- Loss of sensation in an arm or leg, or weakness or paralysis on one side of the body
- Difficulty thinking or speaking, having trouble finding words
- Memory loss, confusion, or disorientation
- Seizures or convulsions
- Personality changes
- Hearing loss
- Difficulty walking normally, losing one's balance, frequent falls
- Weakness and fatigue
- Hydrocephalus

Symptoms of brain tumors in small children may include:

- Increased head size
- In infants, bulging of the soft spot at the top of the skull
- · Breathing problems or problems swallowing

# Diagnosis

Headaches are the single most common symptom of brain tumors in older children and adults, and the one most likely to bring patients to see their doctor—particularly if the headaches are unusually severe or more frequent than those that the person has experienced in the past. Diagnosing a brain tumor requires a series of tests and examinations.

- A family doctor will usually refer the patient to a neurologist, a doctor who specializes in treating disorders of the nervous system. The neurologist will check the patient's balance, vision, hearing, reflexes, coordination, ability to speak clearly, changes in mood, and ability to answer simple questions. These tests help to locate the areas of the brain that might be affected by a tumor.
- Imaging tests. These include computed tomography (CT) scans, magnetic resonance imaging (MRI), and positron emission tomography (PET).
- Electroencephalogram (EEG). This is a test that shows the levels of electrical activity in different parts of the brain.
- Blood and urine tests. These are used to monitor the patient's response to treatment for the tumor.
- Tests for cancer in other organs, since most brain cancers are metastatic rather than primary tumors.
- Biopsy. A biopsy is a procedure in which a sample of tissue is removed for study under a microscope. For a brain tumor, the biopsy may be done as part of an operation to remove the tumor or as a separate procedure.

#### Treatment

Treatment of brain tumors depends on their grade and location. Surgery is usually the first stage. Removing as much of the tumor as possible helps to relieve symptoms as well as slow the growth of the tumor. Some Grade I tumors can be completely removed by surgery.

If the tumor is located in a part of the brain where it cannot be entirely removed by surgery without risk to the brain stem or other parts of the brain that control sight or hearing, or if the tumor is difficult to distinguish from surrounding healthy tissue, the second line of treatment is usually radiation therapy.

Chemotherapy can be given before or after radiation therapy. A newer type of chemotherapy is combined with surgery. After removing as much of the tumor as is possible and safe, the surgeon inserts wafers containing slow-release anticancer drugs.

Treatment for brain tumors may also include rehabilitation therapy for physical aftereffects and supportive therapy to help the patient manage pain and the side effects of cancer treatment.

## Prognosis

The prognosis of a brain tumor depends on whether the tumor is benign or malignant, its location and grade, and the patient's age and overall health. Patients with Grade I tumors that can be entirely removed may recover completely. The prognosis is poorer in very young children and in elderly patients.

The five-year survival rate for patients with malignant primary tumors is less than 10 percent even when these cancers are treated aggressively. Most patients with metastatic brain cancer die as a result of the primary cancer rather than from the metastases in the brain.

#### Prevention

Primary brain tumors are not preventable. Some metastatic brain tumors may be prevented by not smoking, which lowers the risk of lung cancer, or by avoiding overexposure to the sun, which lowers the risk of melanoma.

# The Future

The number of brain tumors diagnosed each year in both adults and children in the United States has been gradually increasing since the 1960s. The reasons for this increase are not yet clear. Theories include toxins in the environment, viruses that have not yet been identified, and heavy long-term use of cell phones.

Some newer treatments for brain tumors that are currently considered experimental include drugs that block the growth of blood vessels in tumors, thus starving them of nutrients. Other new drugs are intended to make radiation therapy more effective by sensitizing the tumor cells to radiation. Gene therapy and biological therapy (which works by stimulating the patient's immune system) are other rapidly expanding areas of research in treating brain tumors.

**SEE ALSO** Alcoholism; Breast cancer; Colorectal cancer; Headache; Lung cancer; Skin cancer

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# WORDS TO KNOW

Benign: Not cancerous.

- **Brain stem:** The lowest part of the brain that connects directly to the spinal cord. It controls such basic life functions as breathing, blood pressure, and heart beat.
- **Cerebellum:** The part of the brain at the lower back of the head just above the brain stem.
- **Glial cells:** Cells in brain tissue that hold nerve cells in place, supply them with oxygen and nutrients, and remove dead nerve cells.
- **Glioma:** A type of brain tumor that starts in the glial cells.
- **Hydrocephalus:** Abnormal buildup of cerebrospinal fluid within the brain.

#### Malignant: Cancerous.

- **Medulloblastoma:** A type of malignant brain tumor that develops in the cerebellum. It is the cancer.
- Meninges (singular, meninx): The protective membranes that cover the brain and spinal cord.
- **Meningioma:** A type of brain tumor that starts in the meninges.
- **Metastasis (plural, metastases):** The spread of a cancer from its original location to other organs or parts of the body.
- **Tumor:** An abnormal mass or growth of tissue that may be either cancerous or noncancerous.

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# U

# Breast Cancer

# Definition

Breast cancer is cancer that starts in the tissues of the human breast. It is the second most common cancer in women, but can also affect men. There are two main types of breast cancer. The more common of the two, ductal carcinoma, begins in the ducts, or tubes, that carry milk from the interior of the breast to the nipple. The other major type is lobular carcinoma. It begins in the lobules, which are the parts of the breast that secrete milk.

# Description

Breast cancer is one of the most feared cancers for women, not only because it is potentially fatal but also because it can lead to disfigurement and worries about the loss of femininity. It can also develop for a long time before obvious symptoms appear. One reason why periodic screening for changes in breast tissue is so important for women over the age of twenty is that breast cancer is highly treatable when caught early.

In the early stages of breast cancer, a woman (or man) may not notice any differences in the size and shape of the breasts. The most noticeable symptom of breast cancer is a lump or thickened area in the breast. Not all such lumps are cancerous; many women notice that the texture of their breasts changes during pregnancy or their menstrual periods. Lumps in the breast can also be caused by noncancerous cysts. Nonetheless, a woman (or man) who notices a lump in the breast should tell their doctor. Also Known As Carcinoma of the breast

#### Cause

Unknown; possibly a combination of genetic factors and hormones

#### Symptoms

Lump in breast; change in size, shape, or feel of breast; fluid leaking from nipple

**Duration** Years

#### **Breast Cancer**

Scanning electronic micrograph of breast cancer cell dividing. © QUEST/SCIENCE PHOTO LIBRARY, PHOTO RESEARCHERS, INC.



# **Demographics**

Breast cancer is one of the most common cancers in women, affecting one in every eight women in the United States in the course of her lifetime. According to the American Cancer Society, there are about 68,000 cases of carcinoma in situ (noninvasive or stage 0 breast cancer) in the United States each year, and 183,000 cases of invasive breast cancer (stages I through IV). About 2,000 American men will be diagnosed with breast cancer.

Breast cancer is the second leading cause of death from cancer among women in the United States; only lung cancer is deadlier. About 41,000 American women and 450 men die each year from breast cancer. The death rate has decreased in recent years, largely as a result of earlier detection. There were an estimated 2.5 million survivors of breast cancer in the United States as of 2008.

Risk factors for breast cancer include:

- Sex. The female/male ratio for breast cancer is 99:1.
- Age. Two out of three invasive breast cancers are found in women over fifty-five.
- Genetic mutations. Between 5 and 10 percent of breast cancers are thought to be caused by inherited mutations in two

# Breast Cancer in Men

Most men think of breast cancer as a disease that affects only women. About one percent of all breast cancers, however, occur in men. About 2,000 men in the United States are diagnosed with breast cancer in an average year and 450 will die from it. Breast cancer is most likely to strike men between the ages of sixty and seventy; the average age at the time of diagnosis is sixty-seven. Men who are most at risk are those with a family history of breast cancer, a mutation in the BRCA2 gene, Klinefelter syndrome, liver disease, exposure to radiation therapy for prostate cancer, obesity, or a history of heavy drinking.

The symptoms, diagnostic tests, staging, and treatment for male breast cancer are similar to those for women. Men have a higher risk of breast cancer spreading to the bones or lungs than women do, however, so early diagnosis and treatment is critical.

genes known as BRCA1 and BRCA2. Women with either of these mutations have an 80 percent chance of developing breast cancer in their lifetime.

- Family history of breast cancer. A woman who has a mother, sister, or daughter with breast cancer has double the risk of developing breast cancer herself.
- Race. Caucasian women are slightly more likely to develop breast cancer than women of other races; however, breast cancers in African American women are often more aggressive. The reason for this difference is not known.
- Early menstruation (before age twelve) or late menopause (after age fifty-five).
- First pregnancy after age thirty or no pregnancy.
- Obesity.
- Smoking.
- Exposure to radiation during adolescence.
- Use of birth control pills or hormone replacement therapy after menopause.
- Heavy drinking.
- History of precancerous changes in the breast.

## **Causes and Symptoms**

The cause of breast cancer is not known. Most researchers think that the disease results from a combination of genetic factors and environmental influences.

Breast cancer has no symptoms in its earliest stages. The first noticeable symptoms may include:

- A lump or thickened area in the breast large enough to be felt during a breast self-examination.
- A watery, bloody, or yellowish discharge from the nipple.
- A change in the shape or size of the breast.
- A flattened, puckered, or indented area in the skin of the breast.
- An orange-peel appearance to the skin of the breast.

#### Diagnosis

Regular screening for breast cancer is important. All women over twenty should learn to perform breast self-examination and check their breasts once a month after the menstrual period. Other screening tests include a breast examination by the doctor as part of a routine office visit, and a mammogram, which is an x-ray study of the breast. Imaging studies (magnetic resonance imaging [MRI] or ultrasound) are done when a mammogram yields abnormal findings.

The definitive test for diagnosing breast cancer is a biopsy. The doctor may remove some tissue through a fine needle (aspiration biopsy) or if a larger sample is needed, through a larger needle (core biopsy). The most accurate technique is a surgical biopsy, in which the surgeon removes all or part of a lump for examination under a microscope.

#### Treatment

The first step in treating any kind of cancer is staging. Staging is a description of the location of the cancer, its size, how far it has penetrated

into healthy tissue, and whether it has spread to other parts of the body. Breast cancer is classified into five stages:

- Stage 0: The cancer is in a lobule or a duct but has not spread beyond it. Breast cancer in this stage is called carcinoma in situ. This type of breast cancer is considered noninvasive.
- Stage I: The cancer is no more than three-quarters of an inch across and has not spread beyond the breast.
- Stage II. The cancer is between three-quarters and two inches across and may have spread to the lymph nodes under the arm.
- Stage III. The cancer has grown into the chest wall or the skin of the breast, is larger than two inches across, and has spread to lymph nodes behind the breastbone and under the arm.
- Stage IV. The cancer has spread to other parts of the body.

Surgery is usually the first line of treatment for breast cancer. Complete removal of the breast and underlying chest muscle, called a radical mastectomy, while more common in the past, is now rarely performed. Surgeons are more likely to recommend one type or another of breastsparing surgery:

- Lumpectomy. In this type of surgery, the surgeon removes the cancer itself and a small amount of tissue around it.
- Partial mastectomy. The surgeon removes the cancer, the breast tissue surrounding it, and some of the underlying muscle.
- Simple mastectomy. The surgeon removes the entire breast.
- Modified radical mastectomy. The surgeon removes the entire breast and nearby lymph nodes but leaves the chest muscles in place.
- Reconstruction. Many women have plastic surgery at the same time as a mastectomy or as a later operation to restore the shape of the original breast. The surgeon may use an artificial implant or the patient's own tissue to reconstruct the breast.

Other treatments for breast cancer that may be used after surgery include:

• Radiation therapy. The radiation may come from a large machine outside the body or from implanted plastic tubes containing a radioactive substance. The tubes remain in place for several days and are removed before the patient leaves the hospital.

- Chemotherapy.
- Hormone therapy. This approach to treatment involves taking drugs by mouth to block the production of estrogen and other female hormones. Estrogen encourages the growth of some breast cancers, and hormone blockers are effective in slowing these tumors in some patients.
- Biological therapy. Also called targeted therapy, this approach stimulates the body's immune system to fight cancer cells rather than attacking them directly. It can also be used to control side effects from chemotherapy and radiation therapy, which often include nausea, vomiting, hair loss, and fatigue.

## Prognosis

The prognosis for breast cancer depends on its stage at the time of diagnosis and the number of lymph nodes that were involved when the cancer was discovered. Women whose tumors were smaller than three-quarters of an inch with no lymph node involvement have a survival rate of 96 percent five years after diagnosis; those with tumors larger than two inches with several lymph nodes involved have a five-year survival rate of only 45 percent.

# Prevention

There are no guarantees that a specific woman will not get breast cancer, but there are some steps women can take to reduce their risk:

- Genetic testing. Women with a family history of breast cancer can get tested for a mutation in the BRCA gene. They can then consult their doctor about their own risk of developing breast cancer.
- Taking drugs that have been shown to reduce the risk of breast cancer. These include tamoxifen and raloxifene.
- Preventive mastectomy. Surgical removal of both breasts before cancer can develop is a treatment sometimes undertaken by women who are at very high risk of breast cancer or who have been diagnosed with a lobular carcinoma in situ.
- Surgical removal of the ovaries. Since the ovaries are the main source of estrogen in a woman's body, this type of operation is sometimes recommended for women with a known BRCA mutation.

Other preventive measures recommended by the American Cancer Society include getting regular exercise, limiting alcohol intake, keeping

# WORDS TO KNOW

**Biological therapy:** An approach to cancer treatment that is intended to strengthen the patient's own immune system rather than attack the cancer cells directly.

**Carcinoma in situ:** A cancer that has not spread or is still in one location in the body.

**Lobule:** One of the glands in the breast that produce milk.

Mastectomy: Surgical removal of the breast.

**Staging:** Measuring the severity or spread of a cancer.

weight at a healthy level, and avoiding the use of hormone replacement therapy after menopause.

# The Future

Researchers are investigating different types or combinations of hormone therapy as a treatment for breast cancer. One large clinical trial is known as the Study of Tamoxifen And Raloxifene or STAR trial. Other research involves new diagnostic techniques for catching breast cancer early.

SEE ALSO Alcoholism; Klinefelter syndrome; Obesity; Prostate cancer

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# **Broken Bones**

See Fractures.

# <mark>m</mark> Bronchitis

# Definition

Bronchitis is a condition in which the tissues lining the bronchi—the two main divisions of the airway that lead into the right and left lungs—become inflamed. Doctors define bronchitis as either acute, a temporary illness that clears up in three weeks or less; or chronic, a recurrent condition in which the person suffers coughing attacks for at least three months over two successive years. Chronic bronchitis is grouped together with emphysema as a form of chronic obstructive pulmonary disease (COPD).

# Description

Bronchitis can be caused by a wide range of disease organisms and other irritants that inflame the tissues of the bronchi, including viruses, bacteria, parasites, smoking, chemical pollutants, or dust particles. Acute

# Also Known As

Chronic obstructive pulmonary disease, COPD

#### Cause

Infections (acute); smoking (chronic)

#### Symptoms

Coughing, wheezing, difficulty breathing, chest pains, fever

#### Duration

A few weeks (acute); years (chronic)

#### **Bronchitis**



Colored x ray of swollen bronchioles (orange) in the lungs of a person with bronchitis. CNRI / PHOTO RESEARCHERS, INC.

bronchitis is caused most often by viruses, while chronic bronchitis is caused most often by smoking.

In acute bronchitis, the tissues lining the bronchi become swollen and irritated by the disease organism or chemical. The air passages slowly become clogged by dead cells and leaking tissue fluid, which further irritates the bronchial tissues. The tissues begin to secrete mucus in response

# When to Call the Doctor about Bronchitis

Patients with acute bronchitis should start to feel better in about two weeks. They should call the doctor if their health does not improve by that time or if they have any of the following symptoms:

- Wheezing and coughing lasts longer than two weeks and is worse at night or during exercise.
- Coughing lasts longer than two weeks and brings up a bad-tasting fluid.
- In addition to coughing, the patients feels very weak and has a fever of 101°F (38.3°C) or higher that will not go down.
- Coughing brings up blood, rust-colored sputum, or yellowish or greenish matter.
- The patient has trouble breathing when lying down.
- The patient's feet swell.

Patients who have repeated episodes of acute bronchitis should see the doctor to be examined for chronic sinusitis or other illnesses. In addition, patients with congestive heart failure, asthma, or emphysema should see the doctor if they develop acute bronchitis, because other lung and heart disorders increase the risk of complications with acute bronchitis. to the inflammation. As the mucus accumulates in the airways, the person starts to cough in order to clear the breathing passages. The coughing, the sputum (mucus and other matter) that is brought up by coughing, and wheezing or shortness of breath are classic symptoms of acute bronchitis. Chest pain may develop after several days of severe coughing.

# Demographics

According to the National Center for Health Statistics, about 14 million Americans have chronic bronchitis. In an average year, there are 12–13 million cases of acute bronchitis reported in the United States, compared to 91 million cases of influenza, 66 million cases of the common cold, and 31 million cases of other acute upper respiratory infections. Acute bronchitis is more common in the winter months in most parts of the United States.

Children are more likely to develop acute bronchitis, while chronic bronchitis is largely a disease of adults. The male/female ratio for chronic bronchitis is about three to two. As far as is known, both acute and chronic bronchitis are equally common in all races and ethnic groups.

Some people are at increased risk of developing bronchitis:

- Smokers and people who live with smokers.
- People with gastroesophageal reflux disease (GERD). GERD is a condition in which stomach acid backs up into the eso-

phagus, which can trigger the coughing reflex.

- People whose jobs expose them to chemicals, dust, or other substances that irritate the airway. These may include cotton and other textiles, wheat and other grains, ammonia, sulfur dioxide, chlorine, and a few other strong chemicals.
- People exposed to high levels of automobile exhaust.
- Infants, elderly people, and others with low resistance to infections.

# **Causes and Symptoms**

The basic cause of bronchitis, whether acute or chronic, is a disease organism or substance that irritates the tissues lining the bronchi.

The symptoms of acute bronchitis are similar to those of a bad cold or other upper respiratory infection:

- Sore throat
- A feeling of tightness or congestion in the chest
- Overall feeling of tiredness
- Low-grade fever and chills
- Difficulty breathing
- Wheezing

# Diagnosis

Acute bronchitis is essentially a diagnosis of exclusion, which means that the doctor must rule out such other illnesses as influenza, strep throat, pneumonia, whooping cough, or tonsillitis. The diagnosis is based on a combination of the patient's history, including a history of recent upper respiratory tract infections or exposure to others with such infections, and a physical examination. During the physical examination, the doctor will listen to the patient's breathing through a stethoscope. A chest x ray may be ordered to rule out pneumonia.

The doctor will sometimes collect a sample of the patient's sputum if there is reason to suspect that the bronchitis is caused by bacteria. Most cases of acute bronchitis are caused by the same types of virus that cause the common cold and cannot be treated with antibiotics; however, bacterial infections can be effectively treated by antibiotic medications. If the patient has a severe sore throat in addition to wheezing and coughing, the doctor may order a rapid strep test to rule out the possibility of strep throat, another type of bacterial infection.

If the patient has had several episodes of acute bronchitis or the doctor has other reasons for suspecting chronic bronchitis, the doctor will order pulmonary function tests (PFTs). These are tests in which the patient is asked to breathe into a device called a spirometer. The spirometer measures how much air the patient's lungs can hold and how fast the air moves in and out of the lungs. It can also be used to determine how well the lungs are exchanging oxygen and carbon dioxide. Another way the spirometer can be used is to test the effectiveness of inhaled medications in treating chronic bronchitis.

#### Treatment

Acute bronchitis is usually treated at home with a combination of bed rest; over-the counter pain-relievers like aspirin, Advil, Tylenol, or Motrin to lower the fever; and over-the counter cough medications. Some of these medications (Robitussin, Mucinex) are intended to loosen the mucus in the bronchial passages while others (Benylin, Pertussin) are intended to suppress (quiet) the coughing. Patients should drink plenty of clear fluids to loosen the mucus and use a humidifier or cool-mist vaporizer to reduce the irritation in the bronchi.

Because the overuse of antibiotics can create drug-resistant organisms, the doctor may not prescribe an antibiotic for acute bronchitis unless he or she has test results indicating that the patient's illness is caused by a bacterium and not a virus.

People with chronic bronchitis may need stronger medications:

- Bronchodilators. Bronchodilators are drugs that work by opening up the airways, which allows for more efficient exchange of carbon dioxide and oxygen. Some are taken in tablet form while others are dispensed in inhalers.
- Steroids. This type of medication works by lowering the inflammation in the tissues lining the airways. These drugs can also be taken in pill form or through inhalers.
- Antibiotics. People with chronic bronchitis may be given antibiotics to lower the risk of developing complications.
- Oxygen. Patients who have severe attacks of chronic bronchitis may need to go to the hospital for oxygen therapy. There are also oxygen tanks that can be used in the home; some of these are portable units.

#### Prognosis

Most cases of acute bronchitis clear up completely in two to three weeks with no long-term complications. Chronic bronchitis, however, increases a person's risk of permanently weakened lungs, heart disease, and a shortened life span.

#### Prevention

The best way to prevent chronic bronchitis is to quit smoking; parents can protect their children from bronchitis by quitting or by not smoking in the first place. In some cases, people whose jobs expose them to

# WORDS TO KNOW

**Bronchus (plural, bronchi):** One of the two major divisions of the airway that lead into the right and left lungs.

- **Diaphragm:** A sheet of muscle tissue that divides the chest cavity from the abdominal cavity.
- **Spirometer:** A device that is used to test the air capacity of a person's lungs and the amount of air that enters and leaves the lungs during breathing.

**Sputum:** Mucus coughed up from the respiratory tract.

chemicals, dust, or other materials that irritate the breathing passage may benefit from changing their occupation.

Acute bronchitis is more difficult to prevent because its most common cause is upper respiratory viruses that are hard to avoid. Regular hand washing and the use of hand sanitizers can reduce the spread of these viruses within a family, school, or day care center. Some people can lower the risk of bronchitis by getting annual flu shots. In addition, people who are over sixty-five or who have diabetes or emphysema may benefit from getting immunized against the most common cause of bacterial pneumonia.

#### The Future

Acute bronchitis is likely to continue to be a commonplace health problem because the viruses that are its most common cause are widespread, particularly during cold and flu season. Chronic bronchitis is expected to affect an even larger proportion of the adult population in the years ahead because many long-term smokers are now reaching the age at which the symptoms of chronic obstructive pulmonary disease are most likely to appear.

**SEE ALSO** Asthma; Common cold; Emphysema; Gastroesophageal reflux disease; Pneumonia; Smoking; Tonsillitus; Whooping cough

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# <mark>m</mark> Bulimia

#### Definition

Bulimia is defined as an eating disorder characterized by recurrent episodes of binge eating followed by self-induced vomiting, laxative or diuretic abuse, vigorous exercise, or fasting.

#### Description

The *Diagnostic and Statistical Manual of Mental Disorders*, fourth edition (DSM-IV), the reference that doctors use to diagnose mental illness, specifies that a person must have an eating binge and try to compensate for it on average twice a week for three months to meet the diagnostic criteria for bulimia. A binge is an episode in which someone consumes a larger amount of food within a limited period of time than most people would eat in similar circumstances. Most bulimics report feelings of loss of control associated with bingeing. A second criterion of bulimia is excessive concern with one's body shape and weight.

There are two subtypes of bulimia, purging and nonpurging, according to the methods used by the patient to prevent gaining weight

Also Known As Bulimia nervosa, BN

#### Cause

Genetic factors, psychological patterns, social influences

#### Symptoms

Binge eating, alternating with dieting, vomiting and/or exercise; laxative abuse

#### Duration

Appears in adolescence or early adulthood; can last a lifetime if not treated

## Princess Diana and Bulimia

Princess Diana (1961-1997) is sometimes credited with bringing bulimia out into the open by talking to the press about her own struggles with the disorder. The princess gave an interview in 1995 in which she described her bulimia: "I had bulimia for a number of years. And that's like a secret disease. You inflict it upon yourself because your self-esteem is at a low ebb, and you don't think you're worthy or valuable. You fill your stomach up four or five times a day-some do it more-and it gives you a feeling of comfort. It's like having a pair of arms around you, but it's temporary, temporary. Then you're disgusted at the bloatedness of your stomach, and then you bring it all up again. . . . It was a symptom of what was going on in my marriage."

When asked by the interviewer whether she had asked for help from anyone else in the royal family, Diana said, "You have to know that when you have bulimia you're very ashamed of yourself and you hate yourself—and people think you're wasting food—so you don't discuss it with people. And the thing about bulimia is your



Diana, Princess of Wales, in 1997. AP IMAGES.

weight always stays the same, whereas with anorexia you visibly shrink. So you can pretend the whole way through. There's no proof."

after a binge. People who have the purging subtype use vomiting, laxatives, enemas, or diuretics to keep from gaining weight; in the nonpurging subtype, the person fasts or overexercises to prevent weight gain. The important point is that bulimics do something after a binge to compensate for their eating. There is another type of eating disorder called binge eating disorder, in which the person has eating binges but does not try to vomit, exercise, or do anything else to prevent gaining weight.

#### **Demographics**

There is some disagreement about the demographics of bulimia, partly because the rules for diagnosing it have changed over time. The usual figure given for bulimia in the United States is 1–3 percent of high school- and college-age women. Many doctors think, however, that

bulimia is underdiagnosed because most people with the disorder are of average weight or only slightly overweight. In addition, there are large numbers of teenagers and young adults who have disordered eating patterns but do not meet the full criteria for bulimia; there may be twice as many young people in this second group as those who meet the full DSM-IV definition.

The gender ratio is usually given as ten females to every one male affected, but some people think that as many as 15 percent of bulimics are male. Gay men appear to be at greater risk of developing bulimia than heterosexual men.

At one time bulimia was thought to affect mostly Caucasian women, but the rates among African American and Hispanic women have risen faster than the rate of bulimia for the female population as a whole, at least in the United States. Occupation appears to be a major risk factor for bulimia. Women whose careers depend on appearance or a certain body build, such as ballet dancers, models, and professional athletes, are reported to be four times as likely to develop bulimia as women in the general population.

#### **Causes and Symptoms**

The causes of bulimia are not known for certain, but are thought to be a combination of genetic factors (possibly unusual sensitivity to foods high in carbohydrates); the emotional climate in the patient's family; and pressures in the wider society to live up to a standardized image of beauty. In terms of family patterns, people with bulimia often describe their families as conflicted and their parents as either distant and uncaring or hostile and critical.

Bulimia is associated with a number of physical symptoms. Binge eating by itself rarely causes serious medical complications, but it is associated with nausea, abdominal bloating and cramping, slowed digestion, and weight gain.

Bulimics who force themselves to vomit after a binge may develop serious medical problems, including:

- Erosion of the enamel on the teeth, caused by stomach acid in the vomited material.
- Enlargement of the salivary glands.
- Scars and calloused areas on the knuckles from contact with the teeth.
- Irritation of the throat and esophagus.
- Low blood pressure and slowed heart rate.

• Electrolyte imbalances. The loss of fluids from repeated vomiting can deplete the body's stores of hydrogen chloride, potassium, sodium, and magnesium. The loss of these chemicals in turn can sometimes affect heart rhythm.

#### Diagnosis

Bulimia is usually diagnosed during an office visit to the patient's primary care doctor, although she or he may be sent to a psychiatrist for an additional evaluation. Primary care doctors are now encouraged to give a screening test to an adolescent or young adult who seems unusually concerned about their weight or asks the doctor a lot of questions about weight loss. These screeners are short sets of five questions about eating habits that the patient can quickly answer. If the doctor thinks that the patient may have bulimia, he or she can look for some of the physical signs that accompany the disorder, such as whether the teeth and salivary glands are normal. In most cases the doctor will order laboratory tests of the patient's blood and urine to make sure that her blood chemistry is normal. Most doctors will also give the patient an electrocardiogram (ECG) to check the patient's heart rhythm. This test is important because some types of chemical imbalances in the blood (from vomiting or using diuretics) can lead to irregular heart rhythms.

Another important part of evaluating a patient for bulimia is a mental status examination. The doctor will need to check the patient for signs of anxiety disorders or depression, because a high proportion of bulimics have a mood disorder. In addition, people with bulimia are more likely to be treated successfully for their eating disorder when their anxiety or depression is also being treated.

#### Treatment

Treatment for bulimia consists of psychotherapy combined with medications. The type of psychotherapy most often recommended for bulimics is cognitive-behavioral therapy (CBT), along with interpersonal therapy. In CBT, the patient is helped to recognize the distortions in their mental image of their body and to correct irrational beliefs about food and eating. Family therapy may be recommended if the patient's family appears to be a major cause of his or her emotional distress; as of late 2007, there was some evidence that family therapy is more helpful for some patients with bulimia than individual therapy. Some bulimics also benefit from group therapy or support group meetings. The medications most often prescribed for bulimics are antidepressants, in particular such drugs as fluoxetine (Prozac) and sertraline (Zoloft). Scientists do not fully understand how these drugs help in treating bulimia, but some think that they help to regulate chemical imbalances in the patient's central nervous system.

#### Prognosis

The prognosis of bulimia depends on several factors, including the patient's age at diagnosis, the quality of family life, and the number of close friendships that she or he has. Patients who are diagnosed early, have good relationships with their parents, and have several close friends are more likely to recover. About half of bulimics have good outcomes after treatment, 18 percent have intermediate outcomes, and 20 percent have poor outcomes.

#### Prevention

While it is difficult to change an entire society and its overly high valuation of physical attractiveness, parents can certainly lower a child's risk of bulimia in later life by creating a warm and loving home. It is important to convey to children that they are loved as whole persons with minds and spirits, not just outwardly pleasing faces and bodies.

#### The Future

It is not known with certainty whether bulimia is increasing in the United States, partly because it overlaps with other eating disorders in some people and partly because doctors are looking more closely at men who may be bulimic but were not diagnosed with the disorder in the past. Although doctors are looking for better treatments of bulimia, including new medications, further research in the chemistry of the brain is needed.

SEE ALSO Anorexia; Depression; Obesity

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# WORDS TO KNOW

Antidepressant: A type of drug given to treat eating disorders as well as mood disorders like anxiety and depression.

**Binge:** An episode of eating in which a person consumes a larger amount of food within a limited period of time than most people would eat in similar circumstances.

**Cognitive-behavioral therapy (CBT):** An approach to therapy that aims at changing distorted thinking patterns, beliefs, and behaviors in order to change the patient's feelings.

**Diuretic:** A type of drug that increases the body's production of urine. Some people with bulimia take diuretics in order to lose weight by reducing the amount of water in the body.

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Something Fishy: Website on Eating Disorders. http://www.something-fishy.org/



# Burns and Scalds

## Definition

Burns are injuries caused to the skin or underlying tissues by direct heat or extreme cold; scalds are burns caused by hot liquids or gases (such as steam).

The severity of burns is measured in two ways: by the depth of the burn (see sidebar) and by the size of the affected area. Doctors use the

#### Burns and Scalds

*Illustration showing first, second, and third degree burns.* THE GALE GROUP.



"rule of nines" to evaluate how much of a person's total body surface area (TBSA) has been burned. In adults, the head and neck together count as 9 percent of the body; each arm also counts as 9 percent. The front of the trunk, the back, and each leg are considered to each represent 18 percent of the TBSA. Doctors count only second- and third-degree burns in estimating the affected areas of the body.

Thermal burns (and scalds) are defined as burns caused directly by heat or by chemical reactions that release heat. They are divided into several categories according to the specific cause:

- Flame burns. This type of burn results from direct contact with fire. Flame burns may take the form of a sudden flash burn produced by a flammable liquid or gas igniting. Pouring lighter fluid on burning charcoal is a common cause of flash burns.
- Contact burns. These burns are caused by touching a hot object like a heated iron or cooking pan. Scalds are contact burns caused by hot liquids or gases.
- Electrical burns. These can be caused by contact with a live electrical wire or in rare cases by lightning. Electrical burns are serious because they can cause serious injury to internal organs that is not obvious from the size of the skin burn.

## Also Known As

Thermal injuries

#### Cause

Exposure to dry or moist heat, radiation, friction, sunlight, extreme cold, electricity, or chemicals

#### Symptoms

Pain, blisters, swelling, redness (minor burns); white or charred, leathery skin (severe burns)

#### Duration

A few days (minor burns) to years (severe burns requiring skin grafting or amputation)

- Radiation burns. Radiation burns can be caused by welding equipment, radiation therapy for cancer, or overexposure to sunlight or tanning equipment. Sunburn is a common form of radiation burn.
- Chemical burns. Chemical burns can be caused by either strongly alkaline materials like lye, or by strong acids like sulfuric acid and nitric acid. Some acids are strong enough to eat through the skin and muscles down to the underlying bone.

Thermal burns affecting a person's airway can be as dangerous as burns affecting the surface of the body. The tissues of the airway can be burned by breathing in superheated air from a fire, toxic gases, smoke, or steam. In the United States, most people who die in house or office fires die from smoke inhalation rather than from skin burns.

Skin can also be burned by exposure to cold, either by lengthy exposure to cold outdoors (frostbite) or by brief exposure to extremely cold materials like dry ice.

# Description

The experience of a burn varies considerably depending on the cause of the burn, the parts of the body that are affected, the severity of the burn, and the situation in which the burn occurred. Human soft tissue will burn at any temperature above 115°F (46°C). Most burns are accidental; however, about 10 percent of cases of child abuse involve intentional burning or scalding of the child.

# **Degrees of Burns**

Burns are classified according to the degree of injury that they cause. Some doctors prefer to classify them according to the layers of skin and muscle tissue that are injured. Both methods of classification are outlined here:

- First-degree burn: Also known as a superficial burn, this type of burn affects only the epidermis, the outermost layer of skin. It produces reddening and minor pain but no blisters.
- Second-degree burn: Also called a partial-thickness burn, a second-degree burn injures the upper level of the dermis, the layer of skin just below the epidermis. Second-degree burns are marked by blisters filled with clear fluid and pain.
- Third-degree burn: Also called a fullthickness burn, a third-degree burn destroys the dermis and some underlying muscle tissue or fascia. The damaged tissue is hard, purplish or white, and there is no pain because the nerve endings in the injured tissue have been destroyed. The hair on the skin is also destroyed in a third-degree burn.
- Fourth-degree burn: In this type of severe burn, the tissue may be destroyed and charred down to the bone. Grafting is necessary in order to save the affected limb.

# **Demographics**

Burns are commonplace injuries worldwide, partly because they have so many potential causes. In the United States, about one million people seek treatment for burns in hospital emergency rooms each year. This figure actually represents a decline from the early 1960s, when the number was about two million. About 40,000 people are hospitalized for burn injuries each year rather than being treated as outpatients; of those admitted to hospitals, 60 percent, or 25,000 people, are sent to one of the 125 hospitals in the United States with specialized burn centers.

According to the American Burn Association, there are about 4,000 deaths from burns each year in the United States, 3,500 from house fires and the remaining 500 from automobile and aircraft crashes, contact with electricity, chemicals or hot liquids and substances. About 75 percent of these deaths occurred at the scene of the accident or on the way to the hospital.

Burn injuries vary according to age group:

- Infants and children: most burn injuries in children four years and younger are caused by scalds (65 percent) and contact burns (20 percent). These injuries often occur when children pull pots and pans containing hot food or boiling water from a stove top.
- Adolescents and children between the ages of four and fourteen: about 88,000 are treated in hospital emergency rooms each year. The most common causes in this age group are hair curlers, curling irons, room heaters, ovens and ranges, clothing irons, gasoline, and fireworks.
- Among adults, men are more likely to require hospitalization for burn injuries than women; 70 percent of patients admitted to specialized burn centers are male. Forty-three percent of these severe injuries occurred at the person's home; 17 percent on the street or highway; 8 percent in the workplace; and 32 percent in other locations.

#### **Causes and Symptoms**

Burns are caused by exposure to a source of heat, whether direct flame, electricity, radiation, contact with a hot object, or chemicals. Airway burns are caused by breathing in steam, heated air from a fire, or smoke and toxic gases.

Symptoms of first-degree burns include pain and reddening of the affected area. Second-degree burns produce blisters as well as reddened skin and pain. Third-degree burns are painless because the nerve endings in the affected area have been destroyed. The affected skin may be white or charred, and is usually dry or leathery. The symptoms of airway burns include:

- · Charred or burned mouth and lips; burns on the face or neck
- Wheezing and difficulty breathing
- · Singed nose hairs or eyebrows
- Coughing
- Dark soot-colored mucus being coughed up
- · Changed voice

## Diagnosis

The diagnosis of burns and scalds is usually obvious from the patient's situation and the appearance of the injury. Patients taken to the emergency room will usually be given a chest x ray to check for evidence of smoke inhalation or other injury to the airway.

Most of the evaluation in the hospital consists of determining the extent of the patient's injuries. The rule of nines is used to estimate the area of body surface covered by second- or third-degree burns. Another quick measurement that can be used to estimate the size of the burn is the area covered by an adult's palm, which is about 1 percent of total body area. Emergency room doctors use these rules to tell whether a patient should be sent to a specialized burn center:

- Third-degree burns covering more than 5 percent of body surface area (BSA)
- Second-degree burns covering more than 10 percent of BSA
- Any second- or third-degree burn on the face, genitals, hands, feet, or skin covering a major joint
- Burns caused by electrical currents, lightning, or chemicals
- Burns occurring together with other major injuries (head trauma, broken bones, etc.)
- Burns in patients with inhalation injuries

#### Treatment

Minor first-degree burns can be treated at home by cooling the injured part by holding it under cool tap water for several minutes. Ice should not be used because it can make the injury to the skin worse. Butter or margarine should not be applied because oily substances will not help heal the burn and may increase the danger of infection. Aspirin or Tylenol can be taken to ease pain, and an antibiotic ointment can be applied to the skin to reduce the risk of infection.

If a person's clothing is on fire, they should be told to "stop, drop, and roll" to put out the fire. They should be pulled away from the hot object or open flame and their clothing removed if possible. Burned clothing that is stuck to the skin should not be removed, however. The burned person should be doused with water and covered with a clean sheet or other cloth while emergency services are called. Severe burns should never be treated with ointments or other household remedies while waiting for emergency help.

Treatment of severe burns in the hospital begins with an evaluation of the patient's airway and their blood circulation. The next step is removal of burned clothing and careful inspection of the burns. After the patient's burns have been washed with sterile solution, he or she will be given intravenous fluids to prevent shock and dehydration. Painkillers are also given intravenously to relieve the patient's pain as quickly as possible.

The next step in burn treatment is surgical debridement (cutting away) of open blisters and dead tissue. If the person's burns are not severe enough to require transfer to a burn center, they will be kept in the hospital overnight to make sure that their airway has not been injured.

#### Prognosis

The prognosis of burns and scalds varies from excellent to poor depending on the location of the burn, its cause, the patient's age and overall health, and how quickly they received treatment. Minor burns rarely cause long-term complications. Second-degree burns, however, can become infected and the infection can spread into the bloodstream if not treated promptly. Patients can also become dehydrated after a burn injury. Anyone with a burn who notices red streaks or pus in the burned area, increased pain, swollen lymph nodes near the burn, or fever; or anyone who feels dizzy, lightheaded, extremely thirsty, or cannot urinate after a burn should see their doctor at once.

According to the American Burn Association, 95 percent of patients treated in specialized burn centers survive their injuries.

#### Prevention

Prevention is one of the most important aspects of burn care, as public health doctors estimate that 75 percent of burns are preventable. The Centers for Disease Control and Prevention (CDC) recommend the following preventive steps:

- Install smoke detectors in the home and check their batteries regularly.
- Teach small children about fire safety, including the dangers of matches and fireworks.
- Set the household water heater at 120°F (49°C) or lower to prevent accidental scalding in the tub or shower.
- Keep small children from climbing up on the stove; turn the handles of pots and frying pans toward the back of the stove to prevent children from grabbing them.
- Place fire extinguishers in key locations in the home and workplace.
- Keep electrical appliances in good repair and keep electrical cords off the floor.
- Practice fire escape routes in the home, school, and workplace.

#### **The Future**

Burns are likely to continue to be a common form of accidental injury. Better education as well as further research into burn treatment may help to lower the number of injuries and deaths caused by burns. One of the most important factors in lowering the rate in the last twenty years is the increased use of smoke detectors. A major new area of research is the development of skin substitutes for covering major burns instead of using grafts taken from the patient's own skin.

SEE ALSO Child abuse; Frostbite; Smoke inhalation; Smoking; Sunburn

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# WORDS TO KNOW

- **Debridement:** The medical term for cutting away dead or damaged tissue from a burn or other injury.
- **Dermis:** The lower layer of skin that contains blood vessels, sweat glands, and hair follicles.

**Epidermis:** The outermost layer of the skin.

- **Fascia:** A sheet of connective tissue that covers and binds together the muscles, glands, blood vessels, and internal organs of the body.
- **Scald:** A burn caused by steam or a hot liquid.

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Genetic

Infection

Unknown

Injury

# m

# Cancer

Cancer is a group of diseases rather than a single disorder. All cancers have three characteristics in common, however. First, they are diseases in which a group of cells begins to grow uncontrollably. Second, the uncontrolled cell growth invades and harms other tissues nearby. Third, cancers metastisize-that is, they spread to other tissues or organs through the lymphatic system or bloodstream. A group of cells can sometimes form a tumor (swelling) that is not cancerous. These benign tumors do not invade other tissues and they do not metastasize but can cause significant illness if growing in a vital area.

Cancer can affect people in any age group; however, the risk for most cancers increases as people get older. Although the diagnosis and treatment of cancer continues to improve, cancers are still responsible for 25 percent of all deaths in the United States and other developed countries. About 7.7 million people die of cancer each year worldwide.

Most cancers are caused by changes in the genetic material of the cells that are multiplying abnormally. These changes may be the result of inherited genetic defects or of such factors as radiation, hormonal imbalances, infectious diseases, or exposure to tobacco smoke and other dangerous chemicals.

SEE ALSO Brain tumors; Breast cancer; Colorectal cancer; Hodgkin disease; Leukemia; Lung cancer; Lymphoma; Prostate cancer; Skin cancer

# **u** Canker Sores

# Definition

Canker sores are small ulcers inside the mouth that are white or grayish with red borders. They are not contagious. Canker sores should not be confused with cold sores, which occur outside the mouth around the lips, are caused by a virus, and are very contagious.

# Description

Canker sores are small patches of raw skin on the tissues that line the cheeks and the inside of the lips. They may also occur at the base of the gums or below the tongue on the floor of the mouth. They may appear as single ulcers or in clusters. Most are about three-eighths of an inch (1 centimeter) wide, but some are larger.

Doctors classify canker sores into three groups: minor sores, which are smaller than half an inch (1.3 centimeters) and heal by themselves in seven to ten days; major sores, which are larger than half an inch (1.3 centimeters), are usually deeper than minor sores, may take as long as a month to heal, and may leave scars; and herpetiform sores, which are small, form clusters that look like the fever blisters caused by herpes viruses, and heal in about a week. About 80–85 percent of canker sores are minor sores, 10 percent are major sores, and 5–10 percent are herpetiform.

#### Also Known As

Aphthous ulcers, aphthous stomatitis, mouth ulcers

#### Cause

Unknown; may be stress-related

#### Symptoms

Small whitish sores inside the lips and cheeks, sometimes under the tongue

**Duration** One to two weeks

# **Demographics**

Canker sores are most common in children over ten years of age and young adults. They are estimated to occur in 30–60 percent of the general population. They appear to be equally common in all races and ethnic groups but are slightly more common in women of childbearing age than in men. Herpetiform sores are more common in older people than in younger patients.

Canker sores appear to run in families even though they are not contagious; about 50 percent of people who have frequent occurrences of canker sores have relatives with the same problem. People with certain digestive disorders, including Crohn disease, irritable bowel syndrome, and celiac disease, are also at increased risk of recurrent canker sores.

#### **Canker Sores**



*Canker sore on the inside cheek.* CUSTOM MEDICAL PHOTO STOCK, INC.

# **Causes and Symptoms**

The exact cause of canker sores is not known; however, some doctors think that the sores develop when the person's immune system targets the tissues of the mouth. There are a number of possible factors that may trigger such a reaction:

- Trauma. Canker sores sometimes develop when a person's mouth is irritated by poorly fitted dentures, loose wires from orthodontic braces, a rough tooth, or accidentally biting the inside of the mouth. Brushing too hard or using a very stiff toothbrush have also been associated with damage to the lining of the mouth and canker sores.
- Nutritional deficiencies. People who are not getting enough vitamin B12, iron, zinc, or folic acid in their diet are more likely to develop canker sores.
- Infection by *Helicobacter pylori*, the same bacterium that causes stomach ulcers.
- Stress. Researchers have noted that high-achieving people and people with higher-than-average anxiety levels are more likely to develop canker sores.
- In women, hormonal changes during menstruation and pregnancy are often associated with an outbreak of canker sores.

# When to Call the Dentist

Canker sores, particularly minor sores, usually heal without difficulty even though they may be painful for a week or so. Patients should, however, see their dentist or doctor if they have any of the following symptoms, which are not normal for ordinary canker sores:

- Fever, particularly fever of 101°F (38.3°C) or higher
- Headache
- Pains in the muscles and joints
- Unusually large sores
- New sores developing before old ones heal
- Sores that extend outward from the lining of the mouth into the lips
- Pain that cannot be controlled by ordinary self-care treatments
- Severe difficulty in eating or drinking
- Nausea, vomiting, or diarrhea
- Sore throat or swollen glands in the neck
- Rash on the face or body

- AIDS and other disorders that affect the immune system.
- Food allergies. Flavoring agents, essential oils, benzoic acid, cinnamon, gluten, cow's milk, coffee, chocolate, potatoes, cheese, figs, nuts, citrus fruits, and certain spices have all been associated with canker sores in some people.
- Sensitivity to certain chemicals found in toothpastes and mouthwashes, particularly sodium lauryl sulfate (SLS), a chemical added to toothpaste to thicken it and create a lather during brushing.

Canker sores are preceded by one or two days of tingling or a mild pain in the area where the sore is developing. The reddened area then turns into a whitish patch of broken skin surrounded by a reddish rim. The area in the middle of the sore that looks white or grey is actually partially destroyed. This is the reason why anything containing acids (fruit juices, sodas, coffee), is hot, or is heavily spiced can cause considerable pain if it touches the sore. Some people find it difficult to brush their teeth, talk, or eat until the sore heals.

#### Diagnosis

The doctor or dentist will usually diagnose canker sores on the basis of their appearance inside the mouth. If the sores do not heal or become more severe, a sample of tissue may be taken to check for other possible mouth disorders. Patients with AIDS, for example, may have an infection in the mouth along with the canker sores. In a very few cases, a sore that does not heal is a symptom of cancer.

# Treatment

Minor canker sores will heal eventually without treatment, although dentists frequently prescribe a mouthwash that contains a steroid medication or a rinse that contains an antibiotic. The antibiotic rinse is not usually given to children because it can cause their teeth to discolor. Other treatments include pastes like Orabase, Aphthasol, or Lidex. These can be applied directly to the sore to speed healing and protect the sore from further irritation by food, tooth brushing, or orthodontic braces. The dentist may also prescribe mouthwashes or gels that contain a local anesthetic. The gels can be applied directly to the sore with a cotton swab.

Patients can also care for canker sores at home by making a solution of 1 teaspoon of salt in a pint of warm water and using it to rinse out the mouth as often as desired. Other home remedies that work for some people include making a paste of baking soda and water to be applied to the sores; allowing small chips of ice to melt slowly over the sores; or applying a small amount of milk of magnesia to the sores several times a day.

Patients with severe canker sores may be treated with steroid medications injected directly into the tissues under the sores. Some dentists have used lasers to treat severe canker sores in patients who do not respond to other forms of treatment. Laser therapy gives good results in treating canker sores but is considered experimental because it requires specialized training to use effectively.

Other treatments that are beneficial for some patients include vitamin and mineral supplements, zinc lozenges, stress management techniques, and avoiding foods that are likely to irritate the mouth. For many people, these "problem foods" include nuts, chips, pretzels, certain spices, salty foods, tomatoes, and citrus fruits.

#### Prognosis

Most canker sores heal on their own in one to two weeks, although major sores may take as long as a month to heal completely. Patients with sores that do not respond to any treatment, take longer than a month to heal, or do not heal at all should see their doctor or dentist as soon as possible.

#### Prevention

There is no way to completely prevent canker sores in people who are susceptible to them because of family history or an underlying disease condition like AIDS. However, maintaining good oral hygiene, eating a nutritious diet, using softer toothbrushes, and checking with the dentist

### WORDS TO KNOW

Aphthous ulcer: The medical term for canker sore. Herpetiform: Resembling blisters caused by herpes. **Stomatitis:** The medical term for an inflammation of the mouth.

to be sure that braces or dentures are fitted properly can all help to lower the risk of canker sores.

#### The Future

Canker sores are a common health problem that is likely to affect about a third of the general population for the foreseeable future. Further advances in laser therapy may offer a new treatment option for people with recurrent or severe canker sores.

**SEE ALSO** AIDS; Allergies; Celiac disease; Cold sore; Crohn disease; Irritable bowel syndrome

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# O Carbon Monoxide Poisoning

## Definition

Carbon monoxide (CO) poisoning is a potentially fatal condition caused by breathing carbon monoxide, which is a colorless and odorless gas produced by the incomplete burning of natural gas, propane, gasoline, kerosene, coal, wood, smoking tobacco, other plant matter, or any other fuel containing carbon. It can also result from breathing the vapors of methylene chloride, a chemical found in paint thinners, degreasers, and solvents.

# Description

Carbon monoxide (CO) is a substance that has always been present in Earth's atmosphere in low concentrations; it can be produced by natural processes as well as by human activity. Most of the naturally occurring CO in the atmosphere comes from volcanic eruptions; however, some is also generated by wildfires. The human body itself produces small quantities of CO as a byproduct of certain metabolic processes.

Carbon monoxide poisoning occurs when fuels or other materials containing carbon are burned without adequate ventilation. The gas is hard to detect without special equipment because it is colorless, odorless, and tasteless. People may not realize they have been exposed until they begin to feel dizzy or headachy, or have trouble breathing.

At concentrations of CO as low as 100 parts per million (ppm), people who are otherwise healthy will feel fatigued, and people with heart conditions will experience chest pains. At higher concentrations, people begin to experience disturbances of vision, loss of memory, nausea, vomiting, confusion, and eventually loss of consciousness and death.

Carbon monoxide has a history as a method of deliberate killing; it was used by the Nazis before and during World War II (1939–1945) to kill people with mental and physical disabilities and some concentration camp prisoners. As of 2008, Illinois, North Carolina, and some other states still permit the use of carbon monoxide chambers to kill unwanted pets.

The Environmental Protection Agency (EPA) stipulates that outdoor concentrations of CO should be no higher than 9 parts per million

#### Also Known As

Carbon monoxide toxicity, CO intoxication

#### Cause

Exposure to carbon monoxide gas

#### Symptoms

Headache, dizziness, nausea and vomiting, chest pain, impaired judgment, unconsciousness, death

#### Duration

Minutes to hours

#### Carbon Monoxide Poisoning

Illustration showing how carbon monoxide enters the blood. © NUCLEUS MEDICAL ART, INC. / ALAMY.



(ppm) for periods of eight hours or longer, or 35 parts per million for one hour. There are no official standards for indoor air. For purposes of comparison, here are some typical levels of carbon monoxide in various locations:

- 0.1 ppm: Normal level of CO in Earth's atmosphere.
- 0.5–5 ppm: Average level in homes without gas stoves.
- 5–15 ppm: Average level in homes with properly installed gas stoves.
- 200 ppm: Average level of automobile exhaust in downtown Mexico City.
- 5,000 ppm: Concentration in the chimney of a fireplace when a wood fire is burning.
- 7,000 ppm: Undiluted exhaust from an automobile without a catalytic converter.
- 30,000 ppm: Undiluted cigarette smoke.

# **Demographics**

The CDC estimates that there are about 2,500 deaths each year in the United States attributed to CO poisoning. About 2,000 of these are suicides, a few are murders, and 500 are unintentional. The highest rates of unintentional poisonings are in cold or mountainous parts of the

country. Many of the unintentional deaths involve automobile exhaust; other leading causes of unintentional deaths are coal, wood, or kerosene stoves and fireplaces; burning of natural gas leaking from a pipeline; combustion of gasoline, acetylene, or utility gas; and industrial sources.

Some people are more susceptible to CO poisoning than others. They include:

- People with asthma, bronchitis, or other lung disorders
- Smokers
- People with chronic heart conditions
- Unborn babies
- Elderly people, particularly those over seventy-five

# **Causes and Symptoms**

The basic cause of CO poisoning is loss of oxygen to body tissues. When a person breathes in carbon monoxide, the gas binds to hemoglobin, the red pigment in red blood cells that transports oxygen to body tissues. When the hemoglobin becomes filled with CO, it cannot carry oxygen to other parts of the body. The heart and the central nervous system are particularly vulnerable to loss of their oxygen supply. If the person goes away from the source of the carbon monoxide, it will take about three hours to get rid of half the CO remaining in the body through the lungs. Giving supplemental oxygen cuts the time to thirty minutes to an hour.

Common sources of CO include:

- Fuel-burning space heaters.
- Charcoal grills and hibachis.
- Gas cooking ranges and water heaters.
- Fireplaces.
- Portable generators.

# Safety Tips

The Environmental Protection Agency (EPA) recommends the following safety measures to reduce the risk of exposure to carbon monoxide:

- Keep stoves and other gas appliances properly adjusted and regularly serviced.
- Have a trained technician inspect, clean, and tune up furnaces, flues, and chimneys every year.
- Repair any leaks promptly.
- Be sure that kerosene heaters are operated with the correct fuel.
- Do not idle cars or trucks inside a garage, even with the doors open.
- Do not use charcoal grills or hibachis indoors or in an enclosed garage.
- Install a carbon monoxide detector with a battery backup and test it once a month. If possible, install a detector on each level of the house and especially near bedrooms. CO detectors should not, however, be installed in kitchens. If the CO detector's alarm sounds, leave the house at once and call the fire department or local utility company from a nearby phone.

People who notice the early symptoms of CO poisoning—headache, dizziness, nausea, vomiting, chest pain, confusion, disorientation—should move outside into fresh air and call for emergency medical care.

- Car and truck exhaust. Children riding in the back of enclosed pickup trucks are particularly vulnerable to exhaust fumes.
- Motorboat engines. Swimmers have been overcome by CO when swimming behind motorboats.

The most common symptoms of CO poisoning have already been described. Some people also develop a sunburn-like skin rash. Firefighters or workers who are employed in certain industries such as pulp mills or metal foundries sometimes develop memory problems or psychiatric symptoms as a result of chronic exposure to CO.

#### Diagnosis

The diagnosis of CO poisoning is not always obvious because some of the early symptoms resemble those of the flu or other illnesses caused by viruses. If the person shows signs of smoke inhalation from a fire or can give a history that suggests recent exposure to CO, the doctor will take a sample of blood to be tested for carbon monoxide levels. In some cases memory loss, confusion, impaired judgment, or other psychiatric symptoms are useful diagnostic clues.

# Treatment

Emergency treatment of CO poisoning involves removing the person from the source of the carbon monoxide as quickly as possible and administering 100 percent oxygen through a face mask. If necessary, the person may also be put on an artificial respirator to help them breathe. In severe cases, the patient may be moved to a hyperbaric oxygen (HBO) chamber. Hyperbaric means that the oxygen in the chamber is at twice or three times normal atmospheric pressure. Treatment in an HBO speeds up the removal of CO from the body, sometimes bringing it down is as little as fifteen to twenty minutes.

# Prognosis

The prognosis of CO poisoning depends on a number of factors, including the patient's age, presence of heart or lung disorders, smoking habits, length of exposure to the carbon monoxide, and the concentration of the carbon monoxide that was inhaled. Most people recover completely with proper treatment; however, some suffer lifelong memory loss, difficulty thinking, or other neurological or psychiatric problems.

# WORDS TO KNOW

**Hemoglobin:** The substance in red blood cells that transports oxygen to the cells of the body.

**Hyperbaric oxygen:** Oxygen that is delivered to a patient in a special chamber at two to three times normal atmospheric pressure.

In general, patients who have gone into coma or whose heart has stopped temporarily have poorer outcomes.

#### Prevention

CO poisoning can be prevented by following the safety tips outlined in the sidebar.

#### The Future

Better public education about the dangers of CO poisoning is part of the Centers for Disease Control and Prevention's plan for the future. According to the National Fire Protection Association, 93 percent of American homes have smoke detectors, but only about 15 percent have carbon monoxide detectors. In addition, the CDC believes that most people are unaware of the dangers of CO poisoning while boating or camping. In terms of treatment, a new portable HBO chamber is presently being evaluated for use by firefighters and other emergency personnel. It has been shown to be safe and effective in laboratory conditions, and was being tested in the field as of early 2008.

SEE ALSO Asthma; Smoke inhalation; Smoking

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# **m** Cataracts

# Definition

A cataract is an opaque, discolored, or cloudy area within the lens of the eye. The lens is composed of water and protein fibers and is normally clear. The clouding of vision associated with cataracts is caused by clumping of the protein fibers in the lens.

# Description

A cataract is a clouding or discoloration in the clear lens of the eye that affects vision. Cataracts may develop in one or both eyes but cannot spread from one to the other. There are three basic types of cataracts caused by aging, which are also called senile cataracts. They are classified by their location in the eye.

- Nuclear. This type develops in the nucleus, the central portion of the lens. Nuclear cataracts are likely to darken, turning a portion of the lens yellow or brown; this type of cataract affects a person's perception of color.
- Cortical. Cortical cataracts develop in the cortex, which is the inner portion of the lens between the nucleus and the capsule. They have a distinctive spoke-like or wedge-shaped appearance and often cause problems with glare.
- Posterior capsular. The capsule is the outermost layer of the lens. A posterior capsular cataract develops within this layer at the back of the lens. People with this type of cataract may see halos around lights or have problems with reading.

Cataracts that are not caused by the aging process include:

- Congenital. Some babies are born with this type of cataract, sometimes as a result of the mother's exposure to rubella. They do not always interfere with vision, however.
- Traumatic. These develop after an injury to the eye, often years after the accident.
- Secondary. This type of cataract develops as a result of diseases like diabetes, or following surgery for another eye disorder such as glaucoma or retinal detachment.

Also Known As Lens opacity

#### Cause

Birth defect; aging; exposure to sunlight

#### Symptoms

Cloudy or fuzzy vision, changes in color perception, seeing halos around lights

#### Duration

Years (unless corrected by surgery)
#### Cataracts

*Illustration of the human eye with a cataract.* SHUTTERSTOCK.



# **Demographics**

Cataracts are widespread among older adults in the general population in the United States, although some children are born with cataracts and others develop cataracts in late childhood. About half of people eighty years and older have cataracts or have had surgery to remove cataracts. According to the National Eye Institute, between 300,000 and 400,000 cataracts severe enough to impair vision are diagnosed each year in the United States, and 1.5 million cataract surgeries are performed. Around the world, cataracts due to aging are the single most important cause of vision loss; one-third of cases of blindness in Africa are caused by cataracts.

As far as is known, race is not a factor in the development of cataracts. With regard to sex, different studies have yielded different results as to whether cataracts are more common in women or men.

### **Causes and Symptoms**

Although researchers do not completely understand why the protein fibers in the lens clump together to form cataracts, they have identified several risk factors for cataract development:

- Age over fifty
- A history of smoking

- Diabetes
- High blood pressure or very low blood pressure
- Exposure to sunlight or other forms of radiation
- A family history of cataracts
- Previous surgery for an eye disorder
- History of injury to the eye or inflammatory diseases affecting the eye
- Long-term use of steroid medications

Cataracts develop slowly and painlessly. Over time, people with cataracts may notice the following signs and symptoms:

- Cloudy, foggy, or filmy vision.
- Need for brighter reading light.
- Increased sensitivity to bright lights and glare; this symptom often affects the person's ability to drive comfortably at night.
- Difficulty telling the difference between some colors (usually blues and purples), or seeing colors as faded or yellowish.
- Need for frequent changes in eyeglass or contact lens prescriptions.
- Seeing halos around lights.
- Seeing double in one eye.
- Increased difficulty in perceiving the contrast between the shape of an object and its background.

# Diagnosis

Cataracts are usually diagnosed by a complete eye examination. After the examiner takes a history of the patient's symptoms (including a family history of cataracts and other eye problems), the patient is usually asked to read the letters on an eye chart known as a Snellen chart to test visual acuity, or sharpness of vision. The patient's eye is then dilated with a medication that keeps the pupil wide open, allowing the doctor to see the retina and the optic nerve to make sure that they are not damaged or abnormal.

# **Progress in Cataract Surgery**

Cataract surgery is now one of the safest surgical procedures, but the high rate of success is relatively recent. In the early twentieth century, cataract removal was a dreaded procedure that all too often had negative effects.

In 1919, after a failed cataract operation on her right eye, American painter Mary Cassatt (1844–1926) underwent a cataract operation in her left eye, before which she wrote to a friend, "I look forward with horror to utter darkness." After the second surgery, she completely stopped painting. Cassatt's friend and colleague Claude Monet (1840-1926) was reluctant to undergo the surgery because of Cassatt's outcome. By 1905, nuclear cataracts in both eyes had changed his perceptions of colors. Eye drops helped for awhile, so he postponed surgery. He finally agreed to it in 1923. Afterward, Monet was very unhappy, writing to his doctor, "I might have finished the [paintings] which I have to deliver in April and I'm certain now that I won't be able to finish them as I'd have liked. That's the greatest blow ... and it makes me sorry that I ever decided to go ahead with the fatal operation." Monet refused to consider an operation on his left eye.

While the eye is dilated, the doctor uses a device called a slit lamp to examine the various parts of the eye in detail by focusing a beam of light into a very small line or slit, which allows the doctor to determine the size and location of a cataract and to exclude glaucoma or other causes of the patient's symptoms.

The doctor will also usually test the pressure of the fluid inside the eye with a device called a tonometer, which also ensures that the patient does not have glaucoma.

#### Treatment

Some cataracts are small enough or growing slowly enough that they may not require treatment, particularly if they are not affecting a person's quality of life. In other cases a change in eyeglass or contact lens prescription may be all that is needed.

If the cataract is large or opaque enough to interfere with the patient's vision, surgical removal is the only effective treatment. There are no medications that can dissolve or clear up cataracts. The two basic types of cataract removal are called small incision cataract surgery and extracapsular surgery. In small incision surgery, the ophthalmologist makes a small incision along the side of the cornea and inserts a device that uses ultrasound waves to break up the clouded lens, which is then removed by suction. This technique is also called phacoemulsification or simply phaco; it can be completed in as little as thirty minutes.

In extracapsular surgery, used primarily to treat cataracts that are too dense to be broken up by phacoemulsification, the ophthalmologist makes a longer incision in the cornea and removes the lens all in one piece.

In most cases, the ophthalmologist inserts a new artificial lens, called an intraocular lens (IOL), inside the lens capsule after the cataract is removed. Some patients cannot use an IOL because they have another type of eye disorder. They are given a new eyeglass prescription or soft contact lenses.

Cataract removal can be done in an outpatient center for eye surgery or a hospital. Most people do not need to stay overnight, but patients should ask a friend or family member to drive them home and help with home care after outpatient surgery. They will need to use eye drops to reduce inflammation and the risk of infection for a few days, and they will not be allowed to bend too far forward or lift heavy objects for several days while the eye heals.

# WORDS TO KNOW

- **Capsule:** The outermost layer of the lens of the eye.
- **Cornea:** The transparent front part of the eye where light enters the eye.
- **Cortex:** The part of the lens underneath the capsule.
- Nucleus: The innermost part of the lens of the eye.
- **Ophthalmologist:** A doctor who specializes in diagnosing and treating eye disorders and can perform eye surgery.
- **Phacoemulsification:** A technique for removing cataracts by breaking up the lens of the eye with ultrasound waves and removing the pieces of the lens by suction.

- **Retina:** The light-sensitive layer of tissue at the back of the eyeball.
- **Retinal detachment:** A disorder in which the retina pulls away from its underlying tissues at the back of the eye.
- **Senile cataract:** Another term for cataracts caused by the aging process.
- **Snellen chart:** A series of letters arranged in lines on a chart to be viewed from a distance of 20 feet (6.1 meters) used to measure visual acuity (clearness of vision). Dutch ophthalmologist Hermann Snellen invented it in 1862.
- **Tonometer:** An instrument used by an ophthalmologist to measure the pressure of the fluid inside the eye.

# Prognosis

Cataract surgery is successful in improving the patient's vision in 90–95 percent of cases. The chief risks are infection, persistent inflammation, changes in the fluid pressure inside the eye, and an increased risk of retinal detachment—a condition in which the retina separates from the tissues at the back of the eye. In a few cases the patient's IOL will need to be removed or replaced. Serious complications occur in fewer than one per 1,000 cataract surgeries.

# Prevention

Cataracts cannot always be completely prevented, but their development can be slowed by a combination of lifestyle changes and good eye care:

- Quitting smoking and using alcohol only in moderation.
- Protecting the eyes by using sunglasses or regular glasses designed to screen out ultraviolet light.

- Eating a balanced diet rich in fruits and vegetables. Some researchers think that the vitamins in these foods may slow the growth of cataracts, although this theory has not been proven.
- Following the doctor's recommendations for such other conditions or diseases as high blood pressure and diabetes.

# The Future

Current research on cataracts includes the possibility of developing medications to treat cataracts. Other research being conducted by the National Eye Institute concerns genetic studies that may help doctors understand how cataracts develop and studies of the effects of sunlight exposure on the lens of the eye.

SEE ALSO Glaucoma; Rubella

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# Cavities

See Tooth decay.



# Celiac Disease

# Definition

Celiac disease is a digestive disorder that runs in families and interferes with the digestion and absorption of the nutrients in foods. It is triggered by an immune reaction to gluten, a protein found in wheat, barley, rye, and triticale (a grain that is a cross between wheat and rye).

# Description

When a person with celiac disease eats foods containing gluten, the tissues that line the walls of the intestine become inflamed because of an immune reaction. The tissues gradually lose their villi, which are small finger-shaped projections that increase the surface area and hence the absorptive capacity of the intestinal wall. Over time the patient may develop some symptoms in the digestive tract, some symptoms elsewhere in the body, or have no symptoms at all until the body's failure to absorb nutrients from the food leads to such conditions as anemia, osteoporosis, or malnutrition.

Although the classic symptoms of celiac disease are failure to thrive in children and diarrhea in adults, so many patients have atypical Also Known As

Celiac sprue, nontropical sprue, gluten sensitivity

#### Cause

Immune reaction to gluten, a protein found in grains

#### Symptoms

Smelly diarrhea and fatigue; some patients have no noticeable symptoms

**Duration** May be lifelong



Patient with a rash associated with celiac disease. © MEDICAL-ON-LINE / ALAMY.

symptoms or none at all that the disease is easily missed during medical checkups.

# **Demographics**

Celiac disease runs in families. A person with a first-degree relative (parent, child, or sibling) who has the disease has a 10 percent chance of developing it themselves. Researchers also think that celiac disease is more common than was once thought. In some parts of Europe, particularly Ireland and Italy, celiac disease affects between one in 250 and one in 300 people. In the United States, as many as one in 133 people, or 2 million in the general population, may have celiac disease.

Celiac disease is thought to be more common in persons of European descent than in Africans or Asians; however, some researchers think that the disease is more widespread in Asia, Africa, and South America than was previously thought.

Celiac disease is most likely to be diagnosed in infants between eight and twelve months of age and in adults between twenty

and forty. It appears to be slightly more common in women than in men.

People with Down syndrome or type 1 diabetes are also at increased risk of developing celiac disease.

# **Causes and Symptoms**

The basic cause of celiac disease is inflammation of the tissues lining the small intestine due to an immune reaction to gluten. The villi in the lining of the small intestine gradually flatten out and disappear, leading to an intestinal surface that is not able to absorb enough nutrients from food to foster growth (in children) or prevent weight loss and other complications in adults. It is not known exactly what triggers the onset of celiac disease; it can occur at any age. In adults, celiac disease often appears after a trauma of some kind, such as infection, physical injury, the stress of pregnancy, or surgery.

Some adults have symptoms in the digestive tract with celiac disease that are often mistaken for symptoms of irritable bowel syndrome:

- Diarrhea, often with watery or frothy stools that have a very foul odor and are usually light tan or gray in color
- Large amounts of intestinal gas
- Weight loss
- Fatigue and muscle weakness related to poor absorption of nutrients
- Abdominal cramps

Some symptoms may appear in other parts of the body:

- Anemia
- Loss of bone strength
- Seizures or tingling numbress in the legs from nerve damage
- In women, missed menstrual periods, difficulty getting pregnant, or repeated miscarriages
- Pale sores inside the mouth

• Bone or joint pain

- Tooth discoloration or loss of tooth enamel
- An itchy skin rash known as dermatitis herpetiformis

# The Gluten-Free Diet (source: American Dietetic Association)

People with celiac disease can eat as much plain (without gravy) meat, fish, rice, fruits, and vegetables as they like because these foods do not contain gluten.

Starchy foods that are allowed: Amaranth, arrowroot, buckwheat, corn, flax, Indian rice grass, nuts, potatoes, quinoa, rice, seeds, soy, tapioca, and wild rice.

Grains to avoid: Wheat (including wheat starch, wheat bran, wheat germ, cracked wheat, and hydrolyzed wheat protein, which are often found in processed foods); barley; rye; and triticale, a grain that is a cross between wheat and rye.

Processed foods that may contain wheat, barley, or rye: bouillon cubes, potato chips, candy, cold cuts, hot dogs, Communion wafers, French fries, matzo, rice mixes, sauces, self-basting turkeys, soups, soy sauce, and vegetables packaged in sauces. Fortunately, most of these foods can be obtained in gluten-free forms from various manufacturers.

Researchers still do not fully understand why the symptoms of celiac disease vary so much from one individual to the next. Some think that the range and the severity of the symptoms are influenced by how long the person was breast-fed, how old they were when foods containing gluten were first added to their diet, and the amount of glutencontaining foods that they presently eat.

People with celiac disease are at risk for complications if the disease is not identified and treated:

- Malnutrition leading to stunted growth in children and osteoporosis in adults
- Epilepsy

- Lactose intolerance
- Kidney stones
- Certain types of cancer, particularly lymphoma and intestinal cancer
- An underfunctioning spleen that can be linked to certain serious infections
- Miscarriage of a pregnancy or birth defects in the baby

#### Diagnosis

It is not unusual for people to suffer from celiac disease for as long as ten years before they are diagnosed. One reason for the delay is that some of the symptoms of celiac disease can be easily confused with the symptoms of other digestive disorders, including Crohn disease and irritable bowel syndrome. Another reason for delayed diagnosis is that many doctors are not knowledgeable about the disease and may not suspect it when a patient comes to them for a checkup. Still another reason is that there are not many laboratories with the special equipment needed to test for celiac disease.

There are several laboratory tests available to diagnose celiac disease. One is a blood test to detect certain autoantibodies in the patient's blood. Autoantibodies are proteins that the body forms in reaction against its own tissues. A more definitive diagnostic test involves taking a small piece of tissue from the lining of the patient's small intestine to check for damage to the villi.

It is important for a person scheduled for these diagnostic tests to keep eating foods containing gluten up to the time they take the tests. If the patient stops eating foods with gluten, the test results may be misleading, indicating that the person does not have celiac disease when in fact they do.

#### Treatment

There is no cure for celiac disease. It is managed by keeping one's diet completely free of gluten and products containing gluten. Unlike lactose intolerance, in which patients can often have small amounts of milk or other dairy products without harm, celiac disease is a condition in which the ingestion of even small amounts of gluten can damage the intestines. A few patients with celiac disease know when they have accidentally eaten foods containing a small amount of gluten because they develop a skin rash.

A patient diagnosed with celiac disease should meet with a dietitian, who is a health care professional with special training in nutrition and in

# WORDS TO KNOW

- Amaranth: An herb that produces seeds used as grain in India, Nepal, Mexico, and parts of South America.
- Autoantibody: An antibody formed in reaction against the tissues of the individual producing it.
- **Gluten:** A protein found in certain grains, particularly wheat, barley, and rye.

- **Quinoa:** A plant grown in Peru and Bolivia for its edible seeds. It is high in protein and easy to digest.
- **Triticale:** A grain that is a cross between wheat and rye, first grown in Scotland and Sweden in the nineteenth century.
- Villi (singular, villus): Small finger-like projections along the walls of the small intestine that increase the surface area of the intestinal wall.

planning specialized diets. The dietitian can help the patient plan meals that include favorite "safe" foods, teach the patient how to read labels on foods, and give advice about eating in restaurants and other situations where gluten-free food may not be readily available. Newly diagnosed people and their families may find support groups for celiac disease to be helpful as they learn to adjust to a new way of life that requires constant attention to food.

#### Prognosis

Patients with celiac disease who are careful to avoid gluten have an excellent prognosis. Healing of the intestine begins within days of excluding gluten from the diet. It takes about three to six months for the intestines to heal in children and younger adults and two years for older adults. A few people, however, have what is called unresponsive celiac disease, which means that their intestines do not heal despite a strictly gluten-free diet. People with this condition may need to be evaluated for complications of the disease, treated with steroid medications, and fed intravenously. Unfortunately, this small group of patients has a poor prognosis.

#### Prevention

There is no known way to prevent celiac disease because the factors that trigger the appearance of the disease are not yet understood.

#### The Future

It is likely that improved diagnostic techniques and greater awareness of celiac disease will lead to a wider variety of gluten-free foods for those

who must avoid gluten. At least two dozen cookbooks and other guides to gluten-free nutrition have been published in the United States just since 2002. Perhaps a cure for the disease lies ahead as well.

**SEE ALSO** Crohn disease; Irritable bowel syndrome; Lactose intolerance; Lymphoma; Osteoporosis

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# U

# Cerebral Palsy

# Definition

Cerebral palsy (CP) is the name of a group of disorders of the central nervous system that affect a person's ability to control body movement and coordination. These disorders typically appear in infancy or early childhood. They are not contagious and are not progressive; that is, they do not get worse over time even though the disabilities are permanent.

# Description

CP refers to various injuries to the brain that can occur during pregnancy or at any point during a child's first three years of life. The severity of symptoms varies considerably; some children are only mildly affected while others are almost completely disabled. Although problems with movement and coordination are the defining characteristics of cerebral palsy, 30–50 percent of patients diagnosed with CP are also mentally retarded, and 15–60 percent have epilepsy. Other patients have problems with vision and hearing.

Doctors distinguish four basic types of cerebral palsy according to the nature of the movement disorder involved.

- Spastic: The most common type of CP, spastic cerebral palsy is characterized by stiffness of the muscles. These children may also have scoliosis (an abnormal curvature of the spine) and possibly seizures. About 81 percent of children diagnosed with CP have the spastic form. It may affect only one side of the body, affect both legs, or affect all four limbs.
- Athetoid (also called dyskinetic): The child's movements have a twisting, jerky, or writhing quality. Children with athetoid CP may drool or make strange faces if the facial muscles are affected by the disorder. About 20 percent of children with CP have the athetoid type.
- Ataxic: The child with ataxic CP has difficulty walking because of poor balance, poor depth perception, and loss of coordination. This is the least common type of CP.
- Mixed: The child has a mixture of symptoms that do not correspond neatly to any of the three previous types.

# Demographics

According to the Centers for Disease Control and Prevention (CDC), about twenty-three children in every 10,000 in the United States have cerebral palsy. Each year about 10,000 children are born in the United States with symptoms of CP. The United Cerebral Palsy Foundation estimates that nearly 800,000 children and adults in the United States are living with one or more of the symptoms of the disorder.

Cerebral palsy affects children of all races and ethnic groups; however, boys appear to be at slightly higher risk than girls. There are about 135 boys with cerebral palsy for every 100 girls with the disorder. Also Known As CP, spastic paralysis

#### Cause

Abnormalities in the brain's ability to control movement

#### Symptoms

Stiff or tight muscles; coordination problems; speech, vision, or hearing problems

**Duration** Lifelong

#### **Cerebral Palsy**



Since the 1960s, doctors have identified a number of risk factors for cerebral palsy:

- Premature birth
- Very low birth weight
- Complicated and lengthy childbirth
- Infection in the mother during pregnancy, most often genital herpes, rubella, or toxoplasmosis
- Head injury
- Multiple births (twins or triplets)
- High blood pressure, seizures, mental retardation, or thyroid disorders in the mother
- Exposure to certain toxic chemicals
- Jaundice in the baby

# **Causes and Symptoms**

At one time it was thought that cerebral palsy was caused by difficulties in childbirth that cut off the flow of oxygen to the baby's brain long

enough to cause brain damage. However, birth complications are now thought to account for only 5–10 percent of cases of CP. It now appears that most cases of CP begin before the baby's birth and are the end result of a number of different factors ranging from genetic mutations to infections and trauma.

Doctors have identified four different types of brain injury that may give rise to CP:

- Damage to the white matter of the brain. This is the part of the brain that transmits nerve signals to other parts of the brain and to the rest of the body. It can be damaged before birth by infections in the mother.
- Abnormal development of the various structures in the brain. This type of brain damage is sometimes caused by genetic mutations that affect the development of the central nervous system. It can also occur before birth as a result of fever, infection, or trauma to the mother.

D.J. Gregory, who has cerebral palsy, follows a player at a golf tournament and writes a blog about his experiences. AP IMAGES.

- Bleeding in the brain. A baby can develop bleeding inside the brain or even a stroke before birth if the mother has high blood pressure.
- Damage caused by loss of oxygen to brain tissue. The parts of the baby's brain that control movement and coordination can be damaged by a long period of oxygen deprivation. This can happen if the mother has low blood pressure or if problems with blood flow to the baby occur.

Cerebral palsy is not always obvious at the time of a child's birth. In many cases, the parents first notice developmental delays; that is, the child does not sit, stand, or walk at the normal times for these milestones. In other cases, the child's muscle tone is not normal; his or her limbs may seem either loose and floppy or stiff and rigid. A common pattern is an early period of low muscle tone and floppy limbs followed by a phase of muscular rigidity and tightness. Other early symptoms of CP may include scissors movements of the legs or other abnormal movements, or reflexes that persist long after they disappear in normally developing children.

Other symptoms of CP may include:

- Difficulty nursing or feeding.
- Seizures.
- Irregular breathing patterns.
- Mental retardation.
- Problems with speech, sight, or hearing.
- Peg-shaped rather than normally shaped teeth, and a high rate of dental cavities.
- Limited range of motion in the joints.
- Difficulties with bowel and bladder control.
- Deformities of the arms or legs. Children who are affected by CP on only one side of the body often have smaller or shorter limbs on the affected side.

# Diagnosis

There is no single laboratory or imaging test that can be used to diagnose cerebral palsy. The diagnosis is based on a thorough physical examination of the child and a detailed history of the mother's pregnancy and childbirth. The baby's parents will be asked for a complete medical history of both the mother's and father's families; the mother's medical problems or infections (if any) before and during pregnancy; and a detailed account of the pregnancy, labor, and delivery. The parents will also be asked to describe the baby's early mental and physical development.

Although CP is often present at birth, it is difficult to evaluate during the first six to nine months of the child's life. Most children with cerebral palsy are diagnosed between one and two years of age.

The doctor may order various tests and imaging studies in order to rule out other possible causes of the baby's symptoms.

- Blood and urine tests. These may be done to rule out disorders caused by hormone imbalances or metabolic disturbances.
- Genetic testing. This type of testing may be done to rule out genetic disorders that are associated with delayed development.
- Imaging studies. Computed tomography (CT) scans and magnetic resonance imaging (MRI) are the techniques most commonly used to evaluate a child with CP.
- Electroencephalogram (EEG). An EEG is a record of the child's brain waves made by a machine attached to electrodes placed at various locations on the child's skull. It may be used to evaluate seizures in children with CP.

# Treatment

There is no cure for cerebral palsy. Treatments for children with CP are individualized because the symptoms vary so much from one child to another; they also often change over time as a child grows older. The various types of treatments and therapies that a child with cerebral palsy may need include:

- Rehabilitation and physical therapy. Children with the spastic form of CP may be treated with splints, braces, casts, walkers, and other devices as well as exercise therapy in order to improve the range of motion in their joints.
- Occupational therapy. This form of treatment helps the child to learn skills that he or she will need for self-care, such as feeding, dressing, and grooming.

- Speech and language therapy. This type of therapy is needed for children with hearing problems related to CP as well as those whose throat and facial muscles are affected by the disorder.
- Medications. These are often prescribed to treat seizures, digestive disorders, and muscle cramps or spasms.
- Nutritional therapy. Children with CP often have difficulties with swallowing food or eating balanced diets. A dietitian may be consulted for advice about special diets or tube feeding.
- Vision and hearing aids.
- Surgery. Some children with cerebral palsy develop severe deformities of the spine or contractures of the muscles that require surgical correction.
- Special education programs. Children with CP range from those with normal intelligence who can often participate in mainstream school programs to those with severe mental retardation. In many cases personal computers can be used by children with CP to improve their communication skills as well as complete homework assignments and keep up with schoolwork.

# Prognosis

The prognosis for people with cerebral palsy depends in part on the severity of their symptoms and the parts of the body that are affected by the brain damage. Those with mild symptoms have a normal life expectancy; however, those with severe problems have a shortened life span. It is estimated that about 25 percent of children with CP have very few limitations on their activities; they can complete school and eventually live independently. Another 50 percent can learn to walk, feed, and clothe themselves, and take care of their body functions but cannot live without some form of part-time help from others. Only 25 percent are so severely disabled that they require extensive care and cannot learn to walk.

# Prevention

Many cases of cerebral palsy cannot be prevented. A pregnant woman can, however, lower her risk of having a child with the disorder by quitting smoking, drinking alcohol, and using drugs during pregnancy—all of these increase the risk of giving birth prematurely. She should also

# WORDS TO KNOW

**Contracture:** A permanent shortening of muscle tissue resulting in deformity or loss of function in a limb or joint.

bile pigments from the liver in the patient's blood.

Jaundice: A yellowish discoloration of the skin and whites of the eyes caused by increased levels of

**Scoliosis:** Abnormal curvature of the spine from side to side.

be tested for immunity to rubella before becoming pregnant and get immunized against the disease if she is not already immune.

#### The Future

The number of people in the United States affected by cerebral palsy has increased since the 1960s. This increase may be due to the fact that larger numbers of premature infants are surviving. Many of these infants will have nervous system defects or suffer brain damage that causes the characteristic symptoms of cerebral palsy. Research is concentrated on ways to detect damage to a baby's brain before birth. Researchers hope that information provided by new approaches to early diagnosis may eventually lead to new treatments for the disorder.

**SEE ALSO** Developmental disability; Genital herpes; Rubella; Scoliosis; Toxoplasmosis

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# Chickenpox

# Definition

Chickenpox or varicella is a highly infectious disease caused by a virus. It is usually a mild disease that used to be common in schoolchildren before 1995, when a vaccine against it was introduced in the United States. It does occasionally produce complications, however, particularly in adults who become infected with it and those with a weakened immune system.

# Description

Chickenpox is a highly contagious disease that spreads rapidly because people can transmit the virus for several days before they feel sick. In most people, the virus enters the body through the nose and throat when the person breathes in air that contains the virus or droplets from an infected person's sneezing or coughing. Once inside the body, the virus multiplies in the lymph nodes of the upper respiratory tract for two to four days after the infection begins. It then enters the bloodstream and travels to the spleen and liver, where it continues to multiply for another week or ten days. At this point the virus invades the skin and begins to produce the skin rash that is a classic sign of chickenpox.

The rash begins as red macules, or flat spots, on the surface of the face and body. Most children have about 250–500 of these spots but may have as many as 1,500. The spots develop in a series of crops or batches that go through characteristic stages. The flat macules grow over a period of ten to twelve hours into papules, or raised reddish pimples. The papules in turn become vesicles, which are fluid-filled blisters that at first look like a dewdrop on a rose petal. The vesicles eventually burst

Also Known As Varicella

**Cause** Varicella-zoster virus

**Symptoms** Itchy skin rash, fatigue, fever, loss of appetite

**Duration** Seven to twelve days



and turn into open sores before crusting over. One of the distinctive marks of chickenpox is that the child will have rashes in several different stages at the same time. The child is contagious until the last of the vesicles has burst and has completely crusted over.

The most common complication of chickenpox is a secondary bacterial infection, which can happen if the child scratches the vesicles and bacteria invade the raw skin underneath. An infection can produce inflammation of the skin and a high fever. Impetigo, another skin disease characterized by sores covered with honey-colored crusts and eventual scarring of the skin, can also result. In some cases bacteria can invade the bloodstream, leading to eventual bacterial pneumonia or infection of the middle ear. Because of the risk of bacterial infection, it is important to keep a child with chickenpox from scratching the skin rash.

Another possible complication is the transmission of the chickenpox virus from a mother who develops the disease to the unborn baby. A

baby whose mother develops chickenpox within five days before delivery or two days after delivery can become very sick from the virus if the mother is not treated.

# **Demographics**

Before 1995, chickenpox was considered one of the classic diseases of childhood, along with measles and mumps; about 4 million cases were reported each year in the United States. About 11,000 people were hospitalized annually with chickenpox in North America, and between fifty and 100 of these people died. Since 1995, the number of cases in the United States has declined about 90 percent, and there are only about ten deaths from chickenpox reported each year—most of them in people who were not immunized. In the United States and Canada, about 90 percent of cases of chickenpox occur in children younger than fourteen years, with the remaining 10 percent affecting older teenagers and adults.

Child with a chickpox rash. © JANINE WIEDEL PHOTOLIBRARY / ALAMY.

There are about 90 million cases of chickenpox reported around the world each year. Whereas chickenpox is most common in children between the ages of one and fourteen years in countries with temperate climates, it affects as many adults as children in countries with tropical climates. In North America chickenpox is most common in late winter and spring.

As far as is known, chickenpox strikes all races and both sexes equally.

### **Causes and Symptoms**

Chickenpox is caused by a virus known as the varicella-zoster virus or VZV. This virus is a member of a family of viruses called human herpesviruses; it is related to the viruses that cause

# Did You Know?

Chickenpox did not get its name because it infects chickens or because humans can get it from eating chickens. The disease was first described in Europe by an Italian doctor named Giovanni Filippo (1510–1580), who gave it its alternate name of varicella. The English word "chickenpox" may come from the fact that the blisters of the skin rash associated with the disease looked like chickpeas to a doctor named Richard Morton, who first wrote about chickenpox in English in the seventeenth century. Chickpeas are sometimes called garbanzo beans; they are edible peas that are often eaten in salads or cooked in stews.

oral and genital herpes infections. The chickenpox virus can be spread by breathing air containing droplets from an infected person's sneezing or coughing, or by direct contact with an infected person or their clothing.

The earliest symptoms of chickenpox occur during its prodrome, a brief period of time that precedes the main symptoms of a disease. During the prodrome, which is more likely to occur in teenagers and adults than in younger children, the person may complain of a head-ache, nausea, general tiredness, and loss of appetite. In many children, the first sign of chickenpox is the appearance of the characteristic rash, accompanied by tiredness and a low-grade fever of 100–102°F (37.8–38.9°C). In a few children the fever may rise as high as 106°F (41.1°C).

The characteristic rash of chickenpox has already been described. It takes about a week for each crop of macules to move through the complete cycle of forming papules, vesicles, and crusted sores. Successive crops of macules usually appear for five to seven days, which means that the child should be considered contagious for at least a week after the appearance of the last group of macules. Fever rarely lasts longer than four days. The child may not feel like eating if there are vesicles forming inside the mouth or throat; the sick child should, however, be encouraged to drink plenty of fluids to prevent dehydration.

#### Diagnosis

The doctor usually diagnoses chickenpox on the basis of its external signs and symptoms; in most cases it is not necessary to take a sample of blood or fluid from the vesicles for a laboratory test. In many cases the doctor is helped in diagnosing the disease by asking whether the child has been exposed to anyone with chickenpox in the last ten to twenty-one days.

It is possible to identify the chickenpox virus in samples of fluid taken from the patient's vesicles, but these tests are expensive, some are slow and may not be reliable, and others require special laboratory equipment that is not readily available to most doctors.

### Treatment

Chickenpox is considered a self-limiting disease; that is, people usually recover from it without requiring special treatments. Children with chickenpox should be kept home from school or daycare to avoid infecting others; however, they do not need bed rest. They should stay home until all the chickenpox blisters have completely crusted over.

The child's fingernails should be trimmed short to prevent the blisters from becoming infected if the child scratches them. Very young children may need to have their hands covered with mittens to reduce the risk of infection from scratching. For relief from the itching caused by the skin rash, children can be bathed in cool water with baking soda added every three to five hours. Other treatments for the itching include Aveeno oatmeal baths, application of calamine lotion, or oral antihistamines.

Children can be given acetaminophen or ibuprofen to bring down the fever and to relieve the headache and sore muscles that sometimes accompany chickenpox. They should never be given aspirin, however, because of the risk of Reye syndrome. Reye syndrome is a two-stage illness of young children that follows a viral infection with fever like chickenpox or flu and is thought to be linked to taking aspirin for the fever.

People with weakened immune systems, such as those with HIV infection or leukemia, should be treated with acyclovir (Zovirax), which is a drug that is usually given to treat oral or genital herpes. Acyclovir can reduce the time it takes for a person with a weak immune system to recover from chickenpox. In addition, adults who develop chickenpox should be given acyclovir because they are at greater risk than children of developing complications, such as pneumonia.

# Prognosis

Most children with chickenpox recover completely; however, some develop skin infections as a result of scratching the blisters, and about 5 percent may develop earaches or pneumonia if the bacteria infecting the skin sores have gotten into the bloodstream. Another common complication is shingles, an inflammation of the skin caused by a flare-up of the chickenpox virus that has remained in the nervous system after the child has recovered from chickenpox itself. Shingles, or herpes zoster, usually occurs decades after the chickenpox episode.

Children with leukemia have a 7 percent mortality rate from chickenpox. In otherwise healthy children, the mortality rate is about two per 100,000 cases.

# Prevention

Most people who recover from chickenpox are immune for their the rest of their lives; it is very unusual for a person who has had chickenpox in childhood to have it again as an adult. Chickenpox can be prevented by immunization with the vaccine introduced in 1995. The vaccine, called Varivax, is known to be effective for at least eight years; however, its longer-term effectiveness needs further study. As of 2008 the American Academy of Pediatrics (AAP) recommended two immunizations with Varivax, the first dose given to infants between twelve and fifteen months of age and the second given to children between the ages of four and six years. Some states have made vaccination for chickenpox mandatory alongside vaccinations for polio, mumps, and measles.

It is important for a woman who is pregnant and has never had chickenpox to check with her doctor about her immune status. Pregnant women or babies younger than one year are not given Varivax. They and people with weak immune systems can be protected against chickenpox by receiving varicella-zoster immune globulin, or VZIG, a preparation made from human immunoglobulin. A pregnant woman who develops chickenpox within five days before her due date or two days after childbirth should be given VZIG and acyclovir to protect the baby's health.

# The Future

Chickenpox is likely to become a less common disease in developed countries if the use of Varivax becomes widespread. The disease is not likely to become extinct, however, because people in developing countries often cannot afford the vaccine.

# WORDS TO KNOW

**Macule:** A spot on the skin or patch that is different in color from normal skin but is usually not raised up above the skin surface.

**Papule:** A small cone-shaped pimple or elevation of the skin.

**Prodrome:** A group of warning signs or symptoms that appear before the onset of a disease.

**Shingles:** A skin inflammation caused by reactivation of the chickenpox virus remaining in the nervous system.

Vesicle: A small blister or sac containing fluid.

Although there has been a tendency on the part of some parents in recent years to avoid having children vaccinated for childhood diseases in the belief that the vaccines have too many side effects, doctors now warn against this attitude.

SEE ALSO Genital herpes; Reye syndrome

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# **m** Child Abuse

# Definition

Child abuse refers to the maltreatment of a child, which can include verbal or physical violence, neglect, or both. Most doctors define four major types of child abuse:

- Physical abuse: Physical abuse includes not only direct physical attacks on a child (hitting, shaking, burning, striking with an object) but also failing or refusing to protect a child from physical injury.
- Emotional abuse: Emotional abuse includes repeated verbal shaming or cursing at a child, terrorizing the child, refusing to speak to the child, isolating, or rejecting the child. Some experts include the kidnapping of a child by a parent or other relative during or after a divorce.
- Sexual abuse: Sexual abuse includes exposing a child to pornography or adult sexual behavior as well as fondling, intercourse, or oral-genital contact with a child. Sexual abuse on the part of an adult related to the child by blood or marriage is called incest.
- Neglect: Neglect refers to omitting or refusing to provide the child with food, shelter, clothing, schooling, or medical care.

# Description

Child abuse was called battered child syndrome when it was first identified in the 1950s because repeated physical injuries to children could be documented by x rays and photographs of external injuries. The definition of abuse was expanded over the years to include neglect, emotional abuse, and sexual abuse. The maltreatment of children and teenagers is more widespread than many people realize; while at least 900,000 children are reported to child protective services in an average year as victims of abuse, researchers think that one out of every seven children between the ages of two and seventeen (eight to nine million children) in the United States is coping with some type of abuse or neglect.

The consequences of abuse depend partly on the type of abuse or neglect, the number of perpetrators involved and their relationship to the

#### Also Known As

Battered child syndrome, child maltreatment

#### Cause

Emotional or mental disturbances in the parents

#### Symptoms

Unexplained physical injuries, fear of parents or other adults, selfdestructive behaviors

Duration

Days to years

# The Children's Protector

C. Henry Kempe (1922–1984), a pediatrician and expert on viruses, was the first doctor to call attention on an international level to child abuse. Kempe had grown up in Germany during the early years of the Nazi regime and left his native land as a teenager. After graduating from medical school in 1945, he planned to devote his career to the study of viruses, but was sidetracked by a newfound interest in treating children's illnesses during a residency in pediatrics at Yale.

Kempe became a professor of pediatrics at the medical school of the University of Colorado in Denver. In the early 1960s he began to notice a pattern of repeated injuries in children that he suspected resulted from parental violence. Dr. Kempe and several of his colleagues then surveyed nearly a hundred hospitals across the United States to find out whether other doctors had seen similar cases. In 1962 they published a paper in the Journal of the American Medical Association that has been described as "the single most significant event in creating awareness and exposing the reality of [child] abuse." In 1972 Dr. Kempe opened a foundation for research in child abuse and protection.

child, the child's age at the time of abuse, and whether and when the child or family gets help. Children physically abused in infancy are at high risk of permanent brain damage from shaken baby syndrome, including vision disorders, learning difficulties, and cerebral palsy.

Children abused emotionally at any age are at risk of depression, panic disorder, memory problems, sleep disorders, eating disorders, anxiety disorders, and suicide attempts. One study reported that 80 percent of a sample of young adults who had been emotionally abused as children had developed one or more psychiatric disorders by age twenty.

Childhood abuse also has significant longterm effects on a person's behavior in adult life. Such adults are 1.5 times more likely to abuse drugs or alcohol than those from healthier families. Adults who were abused as children are more likely to be sexually promiscuous, drop out of school, and have problems getting and keeping a job. A National Institute of Justice study done in 2001 indicated that being abused or neglected as a child increased the likelihood of adult criminal behavior by 28 percent and violent crime by 30 percent. Last, the experience of childhood abuse makes it difficult for adult survivors to form lasting and healthy relationships.

# **Demographics**

Most child abuse occurs at the hands of someone the child knows, usually a parent, other relative, caregiver, or neighbor. According to the Centers for Disease Control and Prevention (CDC), about twelve children in every 1,000 in the United States were reported to child protective services as victims of child maltreatment in 2006, the last year for which data are available. Of these 900,000 children, 64 percent were victims of neglect, 17 percent were physically abused, 9 percent were sexually

abused, and 7 percent were emotionally abused. Girls are slightly more likely to be abused than boys; 52 percent of children reported as victims of abuse were girls.

Some other studies have reported higher rates of emotional abuse as high as 75 percent—when emotional abuse is considered as a factor in other forms of abuse or neglect. Of adults who abuse children, the majority are women—58 percent.

Infants are the most likely age group to suffer abuse; the CDC reported that twenty-four out of every 1,000 children below the age of twelve months were abused in 2006, compared to fourteen per 1,000 for children between one and three years of age, thirteen per 1,000 for children between three and seven, eleven per 1,000 for children between eight and fifteen, and six per 1,000 for teenagers sixteen to seventeen years of age.

Race and ethnicity are also factors. In 2006, the rate of child abuse among African Americans was twenty per 1,000 children; for Native Americans, sixteen per 1,000; and for children of mixed race, fifteen per 1,000.

Child abuse and neglect can lead to death. The CDC reported that more than 1,500 children of all ages died in the United States in 2006 as the direct result of abuse and neglect. Seventy-eight percent of these deaths occurred in children below the age of four years.

#### **Causes and Symptoms**

The basic cause of child abuse and neglect is inadequate parenting. Parents can become abusive or neglectful toward their children for a number of different reasons:

- They were abused themselves as children and do not see abusive attitudes and behaviors as abnormal.
- They suffer from mental illness.
- They lack information about the timetable of normal development in children. For example, they may think that a baby should be toilet-trained at three months of age and they become angry when the baby "fails."
- They belong to a culture that emphasizes harsh physical discipline as the way to rear children.
- They lack impulse control.
- They abuse drugs or alcohol.

The signs or symptoms of child abuse or neglect vary from child to child, but the following are typical:

- Physical abuse: The child has frequent and unexplained bruises, broken bones, burns, black eyes, or scars; does not want to go home from school; shrinks away from adults; seems frightened of his or her parents.
- Emotional abuse: The child seems either too mature or too childish for his or her age; is delayed in emotional development; is either extremely submissive or extremely aggressive toward other children; is not emotionally attached to the parents; has attempted suicide.
- Sexual abuse: The child has sudden changes in appetite, nightmares, or bedwetting; is diagnosed with a sexually transmitted disease; knows about or describes sexual behavior in an adult way; has difficulty walking or sitting; does not want to change clothes for or participate in gym class.
- Neglect: The child begs or steals food; is frequently dirty or has severe body odor; does not have clothes suitable for the weather; says that there is no one home to give care.

# Diagnosis

The diagnosis of child abuse can be complicated because some types of abuse do not leave physical evidence. Teachers, police, doctors, dentists, and in some states clergy are required by law to report suspected child abuse to local law enforcement. Although each state sets its own policies for investigation and protective action, in most states the police and child protective services will investigate the situation and if appropriate, remove the child from the home temporarily or permanently. Final decisions about the child's placement are usually made through the state's family court system.

The child will be given a complete physical examination to look for evidence of traumatic injuries. X rays, computed tomography (CT) scans, or other imaging studies are usually ordered to check for evidence of fractures, bleeding in the brain, damage to the eyes or internal organs, and other injuries. The child may be given a blood test to check for sexually transmitted diseases. The doctor will also rule out a few rare bone or blood diseases that can cause the skin to bruise easily or bones to fracture from very minor injuries.

#### Treatment

The treatment of child abuse is complex and often involves long-term psychotherapy for the parents or other perpetrators as well as the child. Most states require some kind of counseling for the parents.

Children who have suffered severe or long-term abuse often need special education programs as well as physical therapy or medical treatment for their physical injuries.

#### Prognosis

The prognosis of recovery from child abuse varies considerably. Some survivors, sometimes called resilient children, are able to cope with physical injuries and painful memories and do very well in adult life. Others resort to drug and alcohol abuse, criminal behavior, risk-taking, and other self-destructive behaviors; they may eventually attempt or commit suicide. Factors that improve a child's chances of recovering from abuse or neglect include caring adults in the extended family or neighborhood who can serve as role models for the child; a community that takes responsibility for preventing child abuse; and high intelligence in the child.

# Prevention

Prevention of child abuse is a long-term process that requires participation by individuals, families, communities, and the health care and legal systems. Some programs that are yielding good results include home visitation programs aimed at reducing violence in families; foster grandparent programs; educating young parents about children's needs and normal developmental patterns; support groups for single or stressed parents; hotlines for reporting child abuse; and public awareness programs.

Other preventive strategies include teaching children to identify abnormal and abusive behaviors and to report them to family members or other appropriate adults. This approach often helps to identify family members with abusive tendencies before serious injury occurs.

#### The Future

Medical researchers have devoted intense attention to child abuse in recent years. As of 2008, the National Institutes of Health (NIH) was supporting 280 separate studies of treatments and prevention strategies for child abuse. The treatments include various forms of psychotherapy for

# WORDS TO KNOW

**Incest:** Sexual activity between closely related persons, often within the immediate family.

**Neglect:** Failing to meet a child's basic needs for food, clothing, shelter, and medical care.

**Perpetrator:** The person who is responsible for the abuse or neglect.

**Resilience:** The capacity to recover from trauma and other stressful situations without lasting damage.

abused children and reducing drug abuse and other self-harming behaviors in adults who had been abused in childhood. Prevention strategies that are being studied include psychotherapy for abusive parents, parent education programs, home visitation programs, and family strengthening programs.

**SEE ALSO** Cerebral palsy; Depression; Developmental disability; Shaken baby syndrome

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# Childhood Obesity

# Definition

Childhood obesity is a condition that develops when children or teenagers take in more food calories than their bodies burn up. The most common assessment of obesity is made by calculating an individual's body mass index (BMI). Although obesity in adults is measured by the body mass index (BMI), which does not take age and sex into account, the Centers for Disease Control and Prevention (CDC) measure overweight and obesity in children and adolescents by percentiles of BMI.

The CDC has compiled growth charts on the basis of BMI for boys and girls at specific ages. The percentile indicates the relative position of the child's BMI number among children of the same sex and age. In screening children for overweight or obesity, the 85th percentile is regarded as an indicator that a child is "at risk" for overweight. A BMI above the 95th percentile is defined as "overweight." The American Obesity Association (AOA) uses the CDC's 95th percentile cutoff as the definition of "obesity," not just "overweight." There are also some researchers who define obesity in children as body weight at least 20 percent higher than a healthy weight for a child of that height, or a body fat percentage above 25 percent in boys or above 32 percent in girls. A child's primary care doctor may use any or all of these standards for evaluating whether an overweight child is obese.

Also Known As Childhood overweight

#### Cause

Genetic, behavioral, and environmental factors

#### Symptoms

BMI over 85th percentile (overweight) or over 95th percentile (obese)

#### Duration

Most likely into adulthood

#### Childhood Obesity

Families participating in a dance exercise program designed to combat childhood obesity, AP IMAGES.



# Description

Childhood obesity, once a rare condition, has become a major public health concern in the United States and other developed countries. It is a serious health problem not only because it virtually guarantees a lifelong struggle with weight when the young person reaches adulthood but also because it leads directly to diseases and disorders once seen only in adults, including asthma, type 2 diabetes, skin rashes, high blood pressure, liver disorders, and high blood cholesterol levels. In addition to physical problems, obese children are also at risk of depression and other psychological problems related to teasing and criticism of their appearance. Depression in turn can lead to difficulties in school and lifelong underachievement.

# **Demographics**

The percentage of overweight and obese children in North America has tripled since the mid-1970s. In 1976 the percentage of children (defined as youngsters between the ages of six and eleven) defined as obese was 7 percent, and the percentage of obese adolescents (ages twelve to nine-teen) was 5 percent. By 1988 11 percent of young people in both age groups were obese, and by 2000 the percentages were 15.3 percent for

children and 15.5 percent for adolescents. Those figures mean that one American child in every six is obese.

Childhood obesity appears to be more common in girls than in boys, but is more obvious in boys because fat in boys tends to accumulate on the chest and stomach rather than being more widely distributed to other parts of the body.

Childhood obesity is more common in African American, Hispanic, and Native American children than in Asian or Caucasian children. It is also more common in children from families with lower family incomes.

# **Causes and Symptoms**

Genetics is one factor influencing childhood obesity that cannot be changed. Having at least one parent who is obese increases a child's risk of obesity throughout life. Researchers disagree, however, on the importance of genetics as a factor in obesity. Some doctors have pointed out that the rapid rise in the rate of childhood obesity within three decades could not be caused by genetic factors alone. One study reported that 41.95 percent of the children in the study with normalweight mothers were obese or overweight while 34.25 percent of children with normal-weight fathers were obese or overweight. Most doctors in the early 2000s regard childhood obesity as the result of a combination of genetic factors and behaviors (food choices, exercise, and eating habits).

A small percentage of overweight children (less than 10 percent) become obese because of metabolic or genetic disorders. These disorders include Cushing syndrome, caused by a tumor in the pituitary gland; Turner syndrome; achondroplasia (dwarfism); disorders of the thyroid gland; and Prader-Willi syndrome, a rare genetic disorder characterized by mental retardation and an abnormally large appetite for food. In a few cases children become obese as a side effect of medications given to treat rheumatoid arthritis and a few other diseases.

Most doctors believe that the most important factors in childhood obesity are:

• Poor food choices. The easy availability of fast foods and junk foods, combined with parents' allowing children to choose their own foods at home instead of eating shared meals with the family, is one reason why many children become obese.

- Lack of exercise. The popularity of computers, video games, and television as leisure-time activities means that the average child is now much less active than a child in the 1970s. Many children spend as much as four hours a day watching television. Researchers studying a group of 133 children in a suburban community discovered that the obese children were 35 percent less active on school days and 65 percent less active on weekends compared to children of normal weight.
- Psychological factors. Some children learn to use food to calm or comfort themselves when they feel lonely or anxious, in some cases by watching their parents eating when they feel stressed. Unfortunately, stress-related eating sets up a vicious circle in which the child's weight gain often leads to further loneliness or greater anxiety, and more eating.
- Sleep deprivation. Children who do not get enough sleep do poorly in school, which in turn can lead to low self-esteem and overeating.
- Social factors. Get-togethers and other family or group activities centered on food (holiday meals and parties, etc.); advertisements in the mass media that encourage overeating or poor food choices; and schools that have cut back on physical education programs.
- Economic factors. Low family income and obesity are often associated because low-income parents may lack the time and resources to make healthful eating habits and exercise a family priority.

# Diagnosis

The diagnosis of childhood obesity may be based on the CDC body mass index tables for children and adolescents or on other measurements. One common test involves measuring the thickness of the skin fold over the triceps muscle on the upper arm, although this measurement may not be accurate unless performed by a trained technician. Another test involves measuring the child's waist circumference at its widest point, usually at or just below the belly button. If the waist measurement is above the 90th percentile for the child's age and sex, the child is at increased risk of type 2 diabetes and the health complications that accompany it.

# Treatment

Treatment of childhood obesity is broad-based and involves the whole family, not just the affected child or teenager. The child's pediatrician can help draw up a treatment plan. Most plans include the following:

- A reasonable weight loss goal for the child, no more than 1–4 pounds (0.5–1.8 kilograms) per month. An overly ambitious goal is likely to lead to failure and making the child discouraged.
- A dietary prescription from the doctor that specifies the total number of calories per day and recommended percentages of calories from fat, protein, and carbohydrates.
- Increasing the child's level of physical activity to twenty to thirty minutes per day in addition to school sports or other physical education activities.
- Nutrition education. This part of treatment usually involves asking the child to keep a food diary and monitor his or her daily food intake as well as learning about what makes a healthful diet and how the body uses food.
- Family involvement. This may involve nutrition counseling for the parents as well as advising them about substituting family outings focused on physical activity rather than television viewing. Several studies have shown that weight management programs involving the entire family are more successful than those aimed only at the overweight child.

# Prognosis

The likelihood that an obese child will grow into an obese adult depends on three major factors: the age at which the child became obese; the severity of the obesity; and the presence of obesity in at least one parent. Overweight in a child under three years of age does not mean that the child will necessarily be obese in adult life unless at least one parent is also obese. After age three, however, the likelihood that obesity will persist into adulthood increases with the age of the child and is higher in children of any age who are severely obese. After an obese child reaches six years of age, the probability that obesity will persist into adult life is greater than 50 percent; moreover, 70 to 80 percent of obese adolescents will remain obese as adults. The presence of obesity in at least one parent increases the risk of obesity in adult life for children at every age.

Obese children have a poor prognosis for good health in adult life. They are at increased risk for a number of serious long-term health problems, including type 2 diabetes, hypertension (high blood pressure), osteoarthritis, heart attack, stroke, and damage to the eyes, heart, and kidneys.

# Prevention

Prevention of childhood obesity is increasingly important. The American Academy of Pediatrics (AAP) makes the following recommendations for parents:

- If possible, breast-feed children rather than bottle feeding them.
- Respect a child's appetite; do not insist that the child finish every feeding or meal.
- Limit the high-calorie and sugary foods kept in the house.
- Provide a nutritious diet with ample fiber from fruits and vegetables, with no more than 30 percent of calories derived from fat.
- Do not use food as a reward or bribe a child to finish a meal by offering sweets.
- Limit the child's television viewing or video games to no more than two hours per day.
- Encourage the child to participate in sports and other activities involving physical exercise (nature walks, dancing classes, hiking, etc.)
- Plan family activities around outdoor walks, bicycling trips, etc.

# The Future

The CDC expects the percentages of obesity in children and teenagers to continue to rise over the next few decades, as it is unlikely that there will be large-scale changes in people's eating habits and food choices. In addition, it is unlikely that the next few years will see any major breakthroughs in the treatment of obesity in adults.

**SEE ALSO** Achondroplasia; Anorexia; Asthma; Bulimia; Diabetes; Hypertension; Sleep apnea

# WORDS TO KNOW

**Body mass index:** An indirect measurement of the amount of body fat. The BMI of adults is calculated in English measurements by multiplying a person's weight in pounds by 703.1, and dividing that number by the person's height in inches squared.

**Cushing syndrome:** A disorder caused by the excess secretion of cortisol by the pituitary gland.

**Pituitary gland:** A pea-sized gland located at the base of the brain behind the nose that secretes growth hormone and other hormones that affect sexual development and the body's response to stress.

**Prader-Willi syndrome:** A rare genetic disorder characterized by mental retardation and an uncontrollable appetite for food.

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# Definition

Chlamydia is a sexually transmitted disease (STD) caused by *Chlamydia trachomatis* bacteria. It is the most commonly reported infectious disease in the United States. Some strains of *C. trachomatis* can also cause trachoma, an infectious eye disease that can lead to blindness.

# Description

C. trachomatis can be transmitted between sexual partners during oral, anal, or vaginal intercourse. It can also be transmitted from an infected mother to her baby during childbirth and cause an eye infection or a type of pneumonia in the newborn. Chlamydia is sometimes called a silent disease, because it may not produce any noticeable symptoms. For women who do feel sick following infection, the most common symptoms are bleeding between menstrual periods, abdominal cramps, pain during intercourse, and a discharge of pus from the vagina. Men may notice inflammation or soreness in their testicles, pain during urination, or a discharge from the penis. One reason why chlamydia is a dangerous disease in spite of the lack of early warning symptoms in many people is that it can lead to long-term complications for men as well as women. Women with untreated chlamydia are at risk of pelvic inflammatory disease (PID), a condition that can cause lifelong infertility. Some infected people are at risk of developing a type of chronic arthritis called Reiter's syndrome.

# Demographics

Chlamydia is the most common sexually transmitted disease in the United States. The Centers for Disease Control and Prevention (CDC) estimates that there are as many as 4 million Americans infected with chlamydia, with 2.8 million new infections each year. It is highly likely that the real numbers are higher because many people with chlamydia—75 percent of infected women and 50 percent of infected men—do not have any noticeable symptoms and are not screened by a doctor. The World Health Organization (WHO) estimates that about 89 million people are infected

Also Known As Chlamydia trachomatis

infection

#### **Cause** Chlamydia trachomatis bacteria

#### Symptoms (Men)

Painful urination, discharge from penis, pain in testicles

#### Symptoms (Women)

Vaginal discharge, pain in abdomen, pain during intercourse

#### Duration

Weeks to months; complications may last for years

#### Chlamydia



*Image of the* Chlamydia *bacteria.* © MEDICAL-ON-LINE / ALAMY.

with chlamydia worldwide, with about 8 million permanently blind as a result of trachoma.

Some groups of people are at greater risk than others of being infected with chlamydia. High-risk groups in the United States include:

- Young adults of either sex
- Adolescent girls between the ages of fifteen and nineteen
- African Americans
- People who have unprotected sex or do not use barrier methods of birth control (condoms or diaphragms)
- Homosexual men, particularly those who are HIV-positive or abuse drugs or alcohol
- People living in large cities
- People with low income or little education
- People who have a large number of sexual partners

# **Causes and Symptoms**

Chlamydia is caused by *C. trachomatis*, a bacterium that lives inside the cells of the tissues that line the genital tract in both men and women. It can also infect the tissues that line the eye. It takes between one and three weeks for the bacterium to produce noticeable symptoms in infected people; as was noted earlier, however, most infected women and

half of infected men do not develop symptoms troublesome enough to send them to a doctor.

Women infected with chlamydia may notice the following symptoms:

- A burning sensation during urination.
- Pain in the abdomen or lower back
- Fever
- An abnormal discharge from the vagina
- Nausea and vomiting
- Pain during intercourse
- Bleeding between periods

Men infected with chlamydia may experience:

- A burning sensation during urination
- Pain or swelling of the testicles
- A discharge from the penis
- Pain, bleeding, or a discharge from the rectum (in homosexual men)

#### Diagnosis

Chlamydia is not always diagnosed promptly because so many people who are infected have no symptoms and may not go to a doctor. In May 2007, the American College of Obstetricians and Gynecologists (ACOG) recommended annual screening for chlamydia of all sexually active women age 25 and younger, as well as other women at high risk for infection who do not have symptoms. In addition, many people who do have symptoms of chlamydia are also infected with HIV, gonorrhea, syphilis, or other STDs. It is now common for doctors to test patients for these other diseases to determine which disease is causing the patient's symptoms.

Chlamydia can be diagnosed in both men and women by a simple urine test. Another test that can be used is a laboratory culture of a smear taken from a woman's cervix (the lower end of the uterus), the opening of the urethra at the tip of a man's penis, or the anus.

#### Treatment

Chlamydia is treated by a course of oral antibiotics, either as a one-time dose or as a series of pills to be taken over a period of five to ten days. People being treated for chlamydia should not have sex for a period of two weeks after treatment to make sure they cannot pass the infection to others.

# WORDS TO KNOW

**Pelvic inflammatory disease (PID):** Inflammation of the uterus, fallopian tubes, and ovaries caused by chlamydia or gonorrhea. It can lead to permanent inability to have children if not treated.

**Reiter's syndrome:** A type of arthritis than can develop in untreated people with chlamydia.

It is characterized by inflammation of the genitals and the eyelids as well as sore and aching joints.

**Trachoma:** An infectious disease of the eye caused by chlamydia bacteria that can lead to blindness if untreated.

# Prognosis

The prognosis for chlamydia when treated promptly is very good; 95 percent of patients are cured with a single course of antibiotic medications. Between 10 and 40 percent of untreated women, however, will develop pelvic inflammatory disease (PID); of those women who are diagnosed with PID, 5 percent will develop a liver disorder. Reiter's syndrome may also be a long-term or recurrent health problem; more than 40 percent of patients diagnosed with it eventually develop vision problems or permanent arthritis, although they can expect to live normal life spans.

#### Prevention

Chlamydia can be prevented in several ways:

- Abstaining from sexual intercourse
- · Not having sex with high-risk partners
- Using latex condoms every time when having sex
- Avoiding the use of drugs and alcohol, which can impair good judgment
- Having potential sexual partners tested or treated for chlamydia before having sexual relations

Up to one-fourth of patients will be reinfected because their partners were not treated.

#### The Future

The most important task in fighting chlamydia is the introduction of better screening methods, particularly for young women; more effective tracing and treatment of infected partners; and honest communication between patients and their doctors. As one Boston physician said in 2007, "Discussing sexual activity is not easy for a lot of people, but being honest with your physician and getting tested are imperative for good health and future fertility."

#### SEE ALSO AIDS; Gonorrhea; Syphilis

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# Chronic Fatigue Syndrome

## Definition

Chronic fatigue syndrome, or CFS, is a disorder of unknown origin that is characterized by unexplained tiredness lasting a year or more and is accompanied by cognitive (thought-related) difficulties as well as headaches, pain in the muscles and joints, and other symptoms.

# Description

CFS is a disorder that affects a number of body systems, ranging from digestive problems, soreness in joints and muscles, and difficulties with thinking and memory to sleeping problems, chest pain, headaches, and extreme tiredness that is not relieved by rest. Although the disorder bears some resemblance to such other disorders as Lyme disease or fibro-myalgia, it includes some symptoms that do not occur in these disorders. The symptoms of CFS vary in severity from patient to patient and may vary over time for an individual patient.

Many cases of CFS start in the winter following an episode of influenza, an influenza-like illness, bronchitis, or other viral-like illness; however, a significant number appear to be triggered by a period of psychological stress. In a few cases, patients reported that their chronic fatigue began after a blood transfusion. Others have said that their symptoms began for no apparent reason.

CFS carries a high cost in time lost from work and social activities for many people. About half of patients diagnosed with CFS either have to reduce their work load or stop working altogether. One set of studies reported that only 42 percent of CFS patients were employed; of those working, 64 percent limited their work because of CFS, and only 19 percent of patients worked full time without limitations. The illness has a high psychological cost as well, since it makes it hard for people to carry out household chores, complete their education, or plan for the future. Many patients also worry about the long-term impact of chronic fatigue syndrome on their marriage or family relationships.

#### **Demographics**

The Centers for Disease Control and Prevention (CDC) estimates that chronic fatigue syndrome affects between 1 and 4 million people in the United States. Most are young or middle-aged adults, with women being affected four times more frequently than men. It is not known for certain, however, whether women are more likely than men to get CFS or whether they are simply more likely to consult a doctor about their symptoms. Exact statistics about the frequency of CFS are difficult to compile because the definitions of the disorder have changed somewhat since the early 1990s.

CFS affects people of all races and ethnic groups. It appears to be as common among African Americans and Hispanics in the United States

#### Also Known As

CFS, chronic fatigue immune deficiency syndrome, post-viral fatigue syndrome

#### Cause

Unknown but may be caused by infectious diseases

#### Symptoms

Pain in muscles and joints, fatigue, memory problems, headaches, throat pain

#### Duration

May last for years

#### Chronic Fatigue Syndrome

Woman undergoing a title table test to diagnose low blood pressure. Treatment of low blood pressure may help some patients with CFS. DAVID PARKER / PHOTO RESEARCHERS, INC.



as among Caucasians. It also appears to be equally common at all levels of education and income.

# **Causes and Symptoms**

The cause or causes of CFS are still being debated. There are five major theories that researchers have proposed about possible causes of the disorder:

• Viruses. At one time it was thought that CFS is caused by Epstein-Barr virus (EBV) or by the viruses that cause upper respiratory tract. It is now believed that there is no single virus that causes chronic fatigue syndrome.

- An immune system disorder. This explanation is now thought unlikely because CFS does not cause tissue damage like lupus or rheumatoid arthritis.
- A central nervous system disorder caused by high levels of emotional stress. Researchers are still looking into some aspects of this theory.
- Nutritional deficiencies. Although some patients do develop food allergies or become intolerant of certain foods, there is no evidence that poor nutrition plays a role in causing CFS.
- Chronic low blood pressure. The CDC is currently researching the effects of drugs to regulate low blood pressure in patients with CFS; some of these patients appear to benefit from these medications.

The CDC lists the following eight symptoms of chronic fatigue syndrome as primary symptoms (essential to diagnosing the disorder), all of which tend to come and go:

- Significant problems with short-term memory or being able to concentrate
- Sore throat
- Soreness in the lymph nodes
- Muscle pain
- Pain in several different joints that is not accompanied by redness or swelling
- Headaches
- The patient is not refreshed after sleep
- Fatigue that lasts longer than 24 hours after any kind of exertion

Other symptoms that are not considered primary symptoms but are reported by about 50 percent of people with chronic fatigue syndrome include:

- Earaches, jaw pain, chronic coughing
- Chills and night sweats
- Nausea, vomiting, abdominal cramps
- Dizziness and problems with balance
- Depression, anxiety, and panic attacks
- Shortness of breath

- Dry eyes and vision problems
- Chest pain
- Diarrhea
- Weight loss or gain
- Allergies or increased sensitivity to light, noise, odors, chemicals, or medications

# Diagnosis

There are no laboratory tests or imaging studies that can be used to diagnose chronic fatigue syndrome. The diagnosis is primarily a diagnosis of exclusion, which means that the doctor begins by ruling out some diseases and disorders that have similar symptoms. These include disorders that cause fatigue, such as sleep apnea or a thyroid disorder; medications that may cause unusual tiredness; alcohol abuse; recurrence of cancer; severe obesity; or a previous diagnosis of depression, schizophrenia, or an eating disorder.

The CDC has defined two major criteria for CFS. The first criterion is that the person must have severe fatigue for a period of at least six months with other possible causes ruled out. The second criterion is having four or more of the primary symptoms listed above.

# Treatment

Treatment of chronic fatigue syndrome is usually multifaceted, consisting of recommendations about regular exercise and the importance of a nourishing diet, together with regular sleeping habits. About 70 percent of patients with CFS reported that they felt better after completing an exercise program that was designed to keep them active without making their fatigue worse. Antidepressant medications may be given to treat the depression, along with medications to treat the patient's allergies. Patients can also take non-aspirin pain relievers like acetaminophen or ibuprofen to relieve headaches or pain in the muscles and joints. There is, however, no single treatment for chronic fatigue syndrome that helps all patients who try it. What works best for most people is drawing up a personalized treatment plan together with their doctor.

Psychotherapy is recommended to help CFS patients cope more effectively with stress and to improve their chances of returning to fulltime work. The type of psychotherapy that has been found to be most

# WORDS TO KNOW

Chronic: Long-term or recurrent.

**Cognitive:** Pertaining to thinking, learning, or memory.

**Diagnosis of exclusion:** A diagnosis that the doctor arrives at by ruling out other diseases one

by one rather than making the diagnosis on the basis of laboratory tests or imaging studies, or other test results.

Fatigue: A feeling of weariness or tiredness after work, exercise, or emotional stress.

useful for patients with chronic fatigue syndrome is cognitive behavioral therapy or CBT. In this form of therapy, patients learn to identify negative thoughts and behaviors that can complicate getting better, and to replace them with positive attitudes. In addition to CBT, support groups are helpful to many patients in learning to cope with the symptoms of the disease and its impact on their lives.

Some alternative and complementary therapies have been found to be effective in relieving the symptoms of chronic fatigue syndrome. These include gentle forms of physical exercise like yoga and t'ai chi as well as meditation, relaxation techniques, deep breathing exercises, and acupuncture.

#### Prognosis

The prognosis for full recovery from chronic fatigue syndrome is not good for the majority of patients. According to the CDC, between 8 percent and 63 percent of patients report some improvement in their symptoms after treatment. The best estimate, however, is that only 5–10 percent of patients make a full recovery.

#### Prevention

There is no known way to prevent chronic fatigue syndrome because its causes are not yet fully understood.

#### The Future

Research into chronic fatigue syndrome is directed toward identifying any genetic factors that may play a role in the disorder. It is likely that CFS may be redefined as a spectrum of related disorders rather than as a single disease with a clearly identifiable cause.

SEE ALSO Fibromyalgia; Lyme disease; Rheumatoid arthritis

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# m

# Chronic Obstructive Pulmonary Disease

Chronic obstructive pulmonary disease, or COPD, is a progressive (gets worse over time) lung disorder of adults that includes emphysema and chronic (long-term) bronchitis. COPD is also called chronic obstructive airway disease (COAD) or chronic obstructive lung disease (COLD). COPD is characterized by a narrowing of the patient's airway that limits the flow of air to the lungs and causes shortness of breath, coughing, and tiredness. Unlike asthma, however, the partial closure of the airway in COPD is not usually reversible.

COPD is the sixth leading cause of death worldwide, according to the World Health Organization. It is expected to become the third leading cause of death by 2020 because of an increase in the number of smokers in most countries. Smoking accounts for 80–90 percent of cases in the United States, where COPD is the fourth leading cause of death as of 2008. Most of the remaining cases are caused by occupational exposure to dust produced in cotton weaving, gold mining, or coal mining, or by air pollution in large cities. Anyone over forty who has trouble breathing, chronic coughing, sputum production, and a history of smoking or other risk factors should be examined by their doctor for COPD.

SEE ALSO Bronchitis; Emphysema; Smoking

# **m** Cleft Lip and Palate

# Definition

Cleft lip and palate are birth defects that affect the shape and function of the upper lip and the roof of the mouth. A cleft is basically an abnormal split or division. A child may have either a cleft lip (CL), a cleft palate (CP), or both (CL/P).

#### Description

Cleft lip is a birth defect that affects only skin tissue rather than underlying bone or cartilage. It can be categorized as either partial (a small gap or indentation in the baby's upper lip) or complete (the split runs from the upper lip up into the nose). It can also be defined as either unilateral, occurring on only one side of the mouth, or bilateral, occurring on both sides of the mouth.

Cleft palate is a defect in which the two plates of bone that ordinarily join to form the hard palate in the front of the roof of the mouth fail to fuse completely during the baby's development before birth. Cleft palate may be either incomplete, a defect in which only the soft palate at the back of the roof of the mouth is split; or it may be complete, when the hard palate as well as the soft palate is divided.

In addition to complete and incomplete cleft palate, there is a third type of defect affecting the roof of the mouth known as a submucous cleft. This is a condition in which a cleft in either the hard or the soft palate is hidden by a layer of tissue. As a result, the cleft may not be discovered until some months after the baby's birth.

## Demographics

In the United States, between one in 500 and one in 550 babies in the general population are born with cleft lip, cleft palate, or both. There are

**Also Known As** Harelip, orofacial cleft

#### Cause

Combination of genetic and environmental factors

#### Symptoms

Visible split in the lip, palate, or both; feeding problems, speech problems

#### Duration

Lifelong unless corrected by surgery

#### Cleft Lip and Palate

A fourteen-month-old boy shown before his cleft lip and palate were repaired (left), and after. AP IMAGES.



considerable differences among racial and ethnic groups, however. The highest rates of these birth defects are among Native Americans and the lowest rates among African Americans:

- Native Americans: 3.74 per 1,000 babies are born with cleft lip or cleft lip and palate
- Asian Americans: 3.36 per 1,000
- Caucasians: 1.86 per 1,000
- Hispanics: 1.04 per 1,000
- African Americans: 0.93 per 1,000

The male/female ratio varies somewhat depending on the type of defect. Boys are almost twice as likely than girls to have either cleft lip or cleft lip and palate, while girls are slightly more likely than boys to have cleft palate.

#### **Causes and Symptoms**

Cleft lip and cleft palate are caused by the failure of two or more lobes of tissue failing to join correctly during the baby's development during pregnancy. There are five lobes of tissue involved in the formation of the baby's face and upper lip: one that grows downward from the top of the head; two that grow inward from the cheeks; and two that grow inward

just below the cheeks to form the chin and lower lip. If two or more of these five lobes of tissue fail to join completely, a cleft lip or even complete facial deformity may result. The palate is formed later than the upper lip; it is composed of two bony plates that grow together as the last step in joining the five lobes of tissue described earlier. The entire complex process of facial and oral formation can be interrupted by a number of genetic and environmental factors.

Some of the causes of cleft lip and palate that have been identified as of 2008 include:

- A gene mutation discovered in 2004 that increases the risk of cleft lip and palate by a factor of three.
- Genetic disorders involving chromosomal abnormalities, including Down syndrome, Edwards syndrome, and Patau syndrome.
- Nutritional deficiencies in the mother, particularly a lack of vitamin A.
- Exposure to lead, pesticides, and other environmental toxins.
- Use of certain prescription medications, particularly drugs given to control seizures.
- Smoking, alcohol abuse, or use of crack cocaine or heroin by the mother.

The symptoms of cleft lip and cleft palate may include feeding difficulties and failure to gain weight, problems learning to speak, a misshapen nose, crooked teeth, and recurrent ear infections, as well as the visible defects in the baby's face and mouth.

#### Diagnosis

Cleft lip and palate can be determined by the doctor at the time of the baby's birth by a careful physical examination of the newborn. Cleft lip can also be diagnosed before birth by ultrasound.

In addition to a physical examination of the baby, the doctor may recommend genetic testing to rule out a genetic cause of the defect or to evaluate the possibility that the parents will have another child with cleft lip or palate.

#### Treatment

Surgery is the primary treatment for cleft lip and palate. In most cases the baby will be evaluated and treated by a team of doctors and other health

care professionals in order to assess his or her need for speech therapy, dental surgery, and other treatments in addition to repair of the upper lip and palate. The treatment of cleft lip and palate is highly individualized.

Surgical repair of the child's facial abnormalities is often carried out in the following order:

- Repair of a cleft lip. This repair is done between birth and three months of age so that the baby can feed as normally as possible as well as grow and gain weight. If the child has a bilateral cleft lip, the surgeon usually repairs one side first and the other side a few weeks later.
- Placement of ear tubes. These are placed around three months of age in babies with a cleft palate to help prevent ear infections.
- Repair of cleft palate. Done between nine and eighteen months of age.
- Orthodontic treatment. Teeth straightening usually begins when the child is around seven years of age and may continue through age eighteen.
- Cosmetic surgery to improve facial appearance. May be done when the child is fifteen through eighteen years of age.

Speech therapy is included in the child's educational program as needed.

Many children with cleft lip or palate also need some form of counseling or psychotherapy, particularly when they reach adolescence. Although preschoolers with a cleft lip or palate do not seem to feel worse about themselves than other children, by the early school years many children begin to withdraw socially or become fearful of being teased or rejected by classmates. In the teen years, many youngsters with speech problems or facial abnormalities related to cleft lip or palate become depressed because their peers usually rate appearance more highly than such personal qualities as humor or intelligence. Strong parental support can make an important difference in a child's ability to cope with social difficulties, as can counseling by a therapist.

# Prognosis

Cleft lip and cleft palate are very treatable conditions. Most of the physical problems associated with cleft lip and palate, such as crooked teeth, speech difficulties, and an increased risk of ear infections can be treated surgically with a high degree of success. The psychological and social

# WORDS TO KNOW

**Congenital:** Present at birth.

Prenatal: Before birth.

**Hard palate:** A thin bony plate located in the front portion of the roof of the mouth.

**Soft palate:** The soft tissue at the back of the roof of the mouth that does not contain bone.

difficulties of children with these birth defects, however, are less easy to resolve.

## Prevention

Some causes of cleft lip and cleft palate cannot be entirely eliminated. Mothers can, however, lower the risk of having a child with these birth defects by quitting smoking, avoiding the use of alcohol and illegal drugs during pregnancy, and taking vitamin supplements during pregnancy.

## The Future

One exciting new development in the field of cleft lip and palate repair is the possibility of operating on the baby before it is born. Advances in surgery that would allow correction of these facial and oral defects before birth would have many advantages. One benefit is that there would be less scarring; surgery performed on a fetus in the womb is known to leave less noticeable scars than surgery performed after birth. In addition, correction of cleft lip and palate before birth might lower the number of orthodontic and other procedures needed after birth.

**SEE ALSO** Clubfoot; Down syndrome; Ear infection; Edwards syndrome; Patau syndrome

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# u Clubfoot

# Definition

Clubfoot is a birth defect in which one or both of the baby's feet are twisted at a sharp angle to the ankle so that the foot and lower leg resemble the head of a golf club. Clubfoot is usually an isolated congenital (present at birth) problem; it does not mean that the baby has a developmental disorder or defective internal organs.

#### Description

Clubfoot is a congenital defect that varies in severity. Some babies have only one affected foot rather than both. About 50 percent of children have so-called flexible clubfoot, which can be corrected without surgery, while those with what doctors define as resistant clubfoot usually need surgery to correct the position of the foot. Resistant clubfoot is often associated with a thin calf and a small, high heel. Clubfoot does not affect a baby's development until it is time for the infant to start walking. It is important to have the child evaluated for corrective treatment before he

#### Also Known As

Congenital talipes equinovarus, CTEV

#### **Cause** Unknown

#### Symptoms

Deformed foot, difficulty walking normally

**Duration** Lifelong if not corrected

#### Clubfoot



*Baby with clubfoot, which can be corrected with surgery.* © MEDICAL-ON-LINE / ALAMY.

or she begins to walk, as children with clubfoot typically start walking on the outside of the foot. If only one foot is affected, the child will develop a limp. If both feet are affected, walking on the outside of the foot will produce a stiff and awkward-looking gait and weaken the muscles of the child's calf.

# **Demographics**

Clubfoot is one of the most common congenital defects, occurring in one in 1,000 babies in the United States and most countries around the world. It is more common in Polynesia, however, occurring in about seventy-five children per 1,000. Between 30 and 50 percent of children with clubfoot are affected in both feet. Boys are twice as likely than girls to be born with clubfoot in the United States. The reason for the gender difference is not known. In terms of ethnic groups in the United States, the rate of clubfoot appears to be higher in Hispanic babies and lower in Asian American babies. As with the gender ratio, the reason for ethnic differences in the frequency of clubfoot is not known. Clubfoot is not clearly a genetic disorder in the strict sense but appears to run in some families. Parents of a child with clubfoot have a 10 percent chance of having a second child with the deformity. People with clubfoot have a 2 percent chance of having a first-degree relative (sibling, parent, or child) with clubfoot.

#### "Mad, Bad, and Dangerous to Know"

Several famous (or infamous) people have suffered from clubfoot, including Claudius, Emperor of Rome from 41 to 54 AD; haddeus Stevens (1792–1868), a Congressional leader of the antislavery movement during the Civil War; and Josef Goebbels (1897–1945), Adolf Hitler's minister of propaganda during World War II.

George Gordon, Lord Byron (1788–1824), was an English nobleman and poet whose last name became a synonym for brooding unhappiness combined with an extravagant and unconventional lifestyle. Born with a clubfoot that was never corrected, he had a lifelong limp. Some of Byron's biographers believe that this, as well as his troubled upbringing, that caused him to take needless risks in his adult life. In the words of one of his many lovers, he became "mad, bad, and dangerous to know." After his unhappy wife divorced him in 1816, Byron left England permanently. He lived in Italy until 1823, when he became involved in Greece's fight for independence from Turkey. Before leading the rebel army to its first attack, however, he fell sick of a fever and died in a small town in central Greece in



*George Gordon, Lord Byron.* COURTESY OF THE LIBRARY OF CONGRESS.

February 1824. The artist who painted Byron on his deathbed was careful to hide the poet's clubfoot under a strategically placed sheet.

# **Causes and Symptoms**

The cause of clubfoot is not known. Various theories about the cause include the mother's use of certain drugs (particularly MDMA or ecstasy, an illegal drug); compression of the child's foot inside the mother's uterus by a lack of amniotic fluid or by fibrous bands that form inside the amniotic sac; and one or more genetic abnormalities that have not yet been identified. Children with Edwards syndrome, a genetic disorder associated with an extra copy of chromosome 18, are often born with clubfoot or so-called rocker-bottom feet.

Clubfoot is usually obvious at the time of the baby's birth. The affected foot is usually turned outward with the heel turned inward; in some cases, the twisting of the foot may be so severe that it looks as if the foot is upside down. If only one foot is affected, it may be as much as half an inch (1.3 centimeters) shorter than the normal foot. In addition, the calf muscles on the affected leg are usually less developed than those on the normal leg. Clubfoot is not painful to the baby, however, even though it looks uncomfortable.

## Diagnosis

The diagnosis of clubfoot is based on the doctor's examination of the baby's feet after birth. While it is possible to detect clubfoot before birth during an ultrasound test, the ultrasound picture does not allow the doctor to determine how severe the deformity is. After the baby is born, the doctor can examine the foot by feeling the bones and soft tissue, measuring various parts of the foot and the leg muscles, and moving the joints in the foot to see how much range of motion is present in the ankle and toes. The office examination is often followed by an x-ray study of the foot to confirm the diagnosis and to provide a baseline measurement for the surgeon in case surgery is needed later on.

## Treatment

At one time surgery was the only available method of treatment for clubfoot. Since the early 2000s, however, several methods of splinting and bracing have been used to treat flexible clubfoot. The best-known method is the Ponseti method, which was developed by a Spanish doctor named Ignacio Ponseti (1914-) in the 1950s but was not widely used until Dr. John Herzenberg used the Internet to tell parents about the Ponseti method in 2000. The Ponseti method uses gentle placement of the baby's foot into the correct position and keeping it there with a plaster cast. The foot is repositioned and placed in a new cast every few weeks for three to six months. The next stage is fitting the child with special shoes attached to a brace that is worn at night for two to three years. The Ponseti method requires parents to actively participate in their child's foot care; they must be sure that the child wears the brace and special shoes for the required length of time each day and that the shoes and brace are correctly applied. Children with resistant clubfoot are usually referred to an orthopedic surgeon (a surgeon who specializes in operations involving the bones and joints) around six to twelve months of age. The foot is x-rayed again so that the surgeon can plan the exact location of the surgery that is required.

At one time the same technique was used on all children with clubfoot regardless of the degree of correction that was needed, but surgical treatment of clubfoot is now highly individualized. The child is placed under general anesthesia and the surgeon lengthens or loosens certain ligaments or tendons so that the foot can develop normally. The surgeon can usually work only on soft tissues if the child is five years of age or younger. Children older than five, however, usually need to have certain bones in the foot reshaped or removed as well as changes made in the soft tissue of the foot.

#### Prognosis

The majority of children with clubfoot can be treated with good or satisfactory results. The Ponseti method is reported to be successful in 89 percent of patients whose feet can be corrected without surgery. Children whose clubfoot must be treated surgically have good results in 81 percent of cases. The chief measure of satisfaction is the range of motion in the child's ankle following surgery, as the ability to move the ankle freely determines how well the foot will function as the child grows.

The degree of deformity in the foot bones at the time of birth is the major factor that affects the success of surgery. Children who are older have better results in surgery than those who are younger than six months because the foot is larger and the surgeon can better determine the size and location of the tissues or bones that need to be corrected.

#### Prevention

There is no known way to prevent clubfoot, because the causes of the deformity are not yet fully understood.

#### The Future

It is not likely that clubfoot will become significantly more common in the future than it is now. However, further refinements of the Ponseti method and further development of surgical techniques for correcting clubfoot should make it possible for most children to benefit from treatment for the condition.

**SEE ALSO** Edwards syndrome

# WORDS TO KNOW

**Congenital:** Present at birth.

Gait: A person's habitual manner of walking.

**Orthopedics:** The branch of surgery that specializes in the treatment of skeletal disorders or injuries.

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# Co



Also Known As Fever blister, oral herpes

#### Cause Virus

#### Symptoms

Painful blisters on the lips or around the mouth; tingling or pain before

**Duration** Seven to ten days



# Cold Sore

# Definition

Cold sores are blisters on the lips or around the mouth caused by herpes simplex virus 1 (HSV-1) and sometimes by herpes simplex virus 2 (HSV-2). Unlike canker sores, which occur inside the mouth, cold sores are contagious.

# Description

Cold sores are painful blisters that develop on the lips or around the mouth after a person has become infected with the herpes simplex virus. The herpes simplex virus 1 (HSV-1) that generally causes cold sores is related to the herpes simplex virus (HSV-2), that typically causes genital herpes but can also cause cold sores.

People who become infected with HSV-1 develop cold sores within twenty days of infection, although some may develop symptoms sooner. The first episode of infection (called the primary infection) may not have any symptoms or may cause two to three weeks of fever and blisters/sores both in and around the mouth. In most cases, the recurrence of symptoms is preceded by a prodrome, or period of warning symptoms before the main phase of the illness. The prodrome of cold sores usually consists of a tingling, itching, or burning sensation that starts one or two days before the blisters appear. The area of Cold sores around the lips. © MEDICAL-ON-LINE / ALAMY.



skin where the blisters will erupt may swell up, turn red, and be sore to the touch.

The sores themselves last for about a week after they erupt. They appear most commonly on the lips or the area of skin between the upper lip and the nose.

The blisters are small and thin-walled, filled with a clear fluid, and become sores after several days. The HSV-1 virus is shed in the fluid from the sores and can be transmitted to other people if they come in contact with the blisters. This is the stage in the development of cold sores when the infection is most contagious. After a few days, the ulcers form a yellow crust that eventually drops off, leaving an area of pinkish skin underneath. There is no permanent scar from a cold sore. People who get cold sores may have one or two recurrences per year, although some have an outbreak every month and some never have relapses.

# Demographics

According to the National Institutes of Health (NIH), about 80 percent of people in the United States are infected with HSV-1. Most acquire the infection as children from contact with oral fluids from an infected person. Infection with the virus is thought to be equally common in both sexes and all races and ethnic groups. People with weakened immune systems, such as patients being treated for cancer or HIV infection, are at increased risk of getting cold sores if they are exposed to HSV-1.

#### **Causes and Symptoms**

The cause of cold sores is usually herpes simplex virus 1 or HSV-1, although occasionally type 2 may cause the process. The virus enters the body through tiny breaks in the tissues lining the mouth, which is one reason it can easily be spread by kissing or by sharing drinking glasses and other food utensils. HSV-1 then lies dormant in the cells of the nervous system until it is activated by stress, an upper respiratory infection, or some other trigger. It then travels back down the nerves to the skin surface, usually in the same area of skin each time. HSV-2 oral infection usually does not have recurrences.

The symptoms of cold sores have already been described. In most cases, people do not need to see the doctor for ordinary cold sores. They should, however, make an appointment if they have any of the following symptoms or conditions:

- They have HIV or any other illness that weakens the immune system.
- The cold sores do not heal on their own by the end of two weeks.
- The patient's eyes feel sore or irritated.
- The cold sores recur frequently.
- The blisters are unusually large or painful.

# Diagnosis

Most people can tell whether they have cold sores by the way they feel and where they appear. If necessary, the patient's doctor can run a blood test to tell whether the person is infected with HSV-1. The virus can also be cultured from the blister fluid or the sore.

# Treatment

There is no permanent cure for HSV-1 infection. After a person is infected with the virus, it hides within nerve cells, making it difficult for the immune system to find and destroy it. HSV-1 remains in the body, so that cold sores can reappear at any time. Recurrences of oral herpes can be triggered by a number of factors, including getting the flu or a cold, not getting enough sleep, having dental work or oral surgery, getting traveler's diarrhea, menstruation, emotional stress, an injury to the mouth or lips, or exposure to the sun for long periods of time. The connection between colds and flu in reactivating HSV-1 is the reason why oral herpes is commonly known as cold sore or fever blister.

The best time to start treating cold sores is during the prodromal stage before the blisters appear. The doctor can prescribe an antiviral medication to shorten the length of the outbreak and reduce discomfort. Other treatments that can be used are topical anesthetics applied directly to the sores, and aspirin, Advil, or Tylenol to bring down fever. Some people also find that ice applied to the blisters helps to relieve discomfort.

# Prognosis

Most cases of cold sores heal without long-term problems; however, HSV-1 can cause an eye infection that may lead to permanent blindness if fluid from the sores gets into the eyes. For this reason it is important for people with cold sores to avoid scratching or squeezing the blisters.

# Prevention

The NIH recommends the following measures to lower the risk of spreading HSV-1 to other parts of the body or to other people, and to lower the frequency of recurrences.

- Avoid kissing or close contact with others while the blisters are present.
- Avoid sharing items that touch the mouth. These include towels, washcloths, lipsticks, lip balms, razors, and toothbrushes as well as drinking glasses and food utensils.
- Keep the hands clean. Wash them frequently and avoid touching the eyes or genital area during an outbreak.
- Try to avoid such common triggers as colds or flu, high stress levels, or being short on sleep.
- Use sunblock on the lips and face when outdoors for long periods of time.

# WORDS TO KNOW

**Prodrome:** A period before the acute phase of a disease when the patient has some characteristic warning symptoms.

**Topical:** Referring to any medication applied directly to the skin or the surface of the body.

• Some people with frequent recurrences of cold sores benefit from an over-the counter dietary supplement called lysine, which is an amino acid (one of the chemical building blocks of proteins). Patients should check with their doctor to see whether lysine might be helpful for them.

## The Future

Cold sores are likely to continue to be a common health problem in the general population. Although researchers are working on vaccines against both HSV-1 and HSV-2, an effective vaccine against either herpes virus is at least several years away.

SEE ALSO Canker sores; Genital herpes

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# Colorectal Cancer

# Definition

Colorectal cancer develops in the colon (the first 4-5 feet [0.6–0.9 meter] of the large intestine) or the rectum (the last few inches of the large intestine).

# Description

Colorectal cancer affects the lower part of the digestive tract. It occurs most often in people over 50 but may develop in younger adults with a family history of colorectal cancer. Most colorectal cancers develop out of polyps, tissue growths that arise out of the tissue that lines the large intestine. Most polyps are benign, but some undergo changes in their genetic makeup that cause them to eventually become cancerous.

Colorectal cancer usually develops over a period of several years. In many cases the patient has no symptoms but is diagnosed as the result of screening for the disease. About half of patients go to their doctor because they have abdominal pain; a third notice a change in bowel habits; and 15 percent have an obstruction (blockage) in their intestines. In some cases these patients may notice that their bowel movements are unusually thin in shape. As a rule, the larger the cancer and the closer it is to the anus, the more likely the patient is to have noticeable changes in bowel habits.

# Also Known As

Colon cancer, cancer of the large bowel

#### Cause

Unknown; genetic in about 3 percent of cases

#### Symptoms

Changes in bowel habits, blood in the stool, abdominal pain, unexplained weight loss

#### Duration

Several years

# **Demographics**

Colorectal cancer is the fourth most common cancer in the United States. According to the American Cancer Society (ACS), about 112,000 people are diagnosed with colon cancer annually; about 41,000 new cases of rectal cancer are diagnosed each year; and about 57,000 persons die each year from colorectal cancer. According to the World Health Organization (WHO) there are about 940,000 new cases of and 500,000 deaths from colorectal cancer reported worldwide each year.

Colorectal cancer is most common in older adults; the average age at the time of diagnosis is 72. Colon cancer in teenagers or young adults is



A virtual colonoscopy produces pictures of the inside of the colon; it is used to screen for colorectal cancer. APHP-PSL / PHOTO RESEARCHERS, INC.

unusual. Rates are equal for men and women. African Americans appear to have higher rates of colon cancer than members of other racial groups in the United States, but the reasons are unclear.

The lifetime risk of developing colon cancer in the United States is about 7 percent. Researchers at the National Cancer Institute have identified several factors that increase a person's risk of colon cancer:

- Age over 50.
- A family history of colorectal cancer. Parents, siblings, or children of a person diagnosed with colon cancer have an increased risk of developing it, particularly if the relative was diagnosed at a young age.
- Having either of two specific genes that increase the risk of colon cancer. These genes are associated with 3 percent of all colon cancers, and can lead to colon cancer by age forty. They can be detected by genetic testing.
- In women, a personal history of breast cancer.
- A history of ulcerative colitis or Crohn's disease.
- A history of polyps in the colon or rectum. Polyps are growths that develop along the inner wall of the colon or rectum, most often in people over fifty. Most are benign (not cancerous), but some may develop into cancerous tumors.

# Good Grief, Charlie Brown!

Charles Schulz (1922–2000), the creator of *Peanuts*, one of the most beloved comic strips of the twentieth century, died of colon cancer only 60 days after being diagnosed with the disease in November 1999. The diagnosis made headlines because Schulz had originally entered the hospital for surgery for another disorder. He announced his retirement from cartooning on December 14 and died in his sleep on February 12, 2000.

Schulz's mother died of cancer when he was a young man, but he apparently was unaware of or ignored his own increased risk of developing the disease, as well as some early warning symptoms. He said in a radio interview shortly after his diagnosis, "I never dreamed that this [cancer] would happen to me. I always had the feeling that I would stay with the strip until I was in my early eighties, or something like that. But all of sudden it's gone. I did not take it away. This has been taken away from me."



*Charles M. Schulz.* COURTESY OF THE LIBRARY OF CONGRESS.

- A diet high in fat and low in fiber.
- A history of heavy smoking or alcohol consumption.
- Obesity and diabetes.
- Gigantism and other disorders involving growth hormone.
- Previous radiation treatment for cancers elsewhere in the abdomen.

# **Causes and Symptoms**

The cause of most cases of colorectal cancer is the change in normally benign intestinal polyps to cancerous tumors. There are several different types of intestinal polyps, but only two carry a risk of developing into cancers. These two types can be removed during screening tests for colorectal cancer. The triggers that cause some polyps to become cancerous are not completely understood. In addition to changes in bowel habits, abdominal cramping, and signs of intestinal blockage, patients with colorectal cancer may have the following symptoms:

- General tiredness, unexplained weight loss, and lack of appetite
- Bleeding from the rectum or blood or mucus on the stools
- Anemia
- Pain when passing a bowel movement
- Nausea and vomiting
- A feeling that the bowel hasn't completely emptied following a bowel movement

# Diagnosis

Doctors may use several methods to screen for colorectal cancer. The simplest are a digital rectal examination (DRE) and a fecal occult blood test (FOBT). In a DRE, the doctor inserts a gloved finger into the lower part of the rectum to feel for tumors. The FOBT requires the patient to take a kit home from the doctor's office and collect a stool sample, which is then returned to the doctor or a laboratory to be tested for occult (hidden) blood. Patients must avoid eating rare meat and other foods that can affect the test results before using the kit.

Other tests that may be used to diagnose colorectal cancer include:

- Blood tests for tumor markers, substances that can be analyzed to detect the presence of cancer.
- Barium enema. Barium in enema form is given to coat the lining of the colon and rectum. Air is then blown into the colon in order to fill it. The resultant x-ray can be used to detect precancerous polyps as well as cancerous tumors.
- Sigmoidoscopy. A sigmoidoscope is a flexible lighted tube that can be inserted into the rectum and used to examine the last 2 feet (0.6 meter) of the colon. It can be done in a doctor's office but does not provide a view of the entire colon. If a polyp or tumor is found, the doctor will recommend a colonoscopy in order to check the upper part of the colon.
- Colonoscopy. A colonoscope is a long flexible tube attached to a video camera and monitor that allows the doctor to examine the entire length of the patient's colon and rectum. The patient must take a laxative the night before to cleanse the bowel and may be

given a sedative in the doctor's office to make them more comfortable. The doctor can remove polyps during a colonoscopy or take tissue samples for analysis.

• Virtual colonoscopy. This technique uses computed tomography (a CT scan) to take images of the patient's colon but is not yet available in all medical centers. Although virtual colonoscopy does not involve inserting a tube into the patient's rectum, the patient must still take a laxative the night before to empty the bowel.

# Treatment

The first step in treating colorectal cancer is called staging. Staging describes the location of the cancer, its size, how far it has penetrated into healthy tissue, and whether it has spread to other parts of the body. Colorectal cancers are classified into five stages:

- Stage 0: The cancer has not grown beyond the lining of the colon or rectum.
- Stage I: The cancer has penetrated through the lining of the colon or rectum into the underlying tissues but has not spread beyond the colon wall.
- Stage II. The cancer has grown through the wall of the colon or rectum but has not yet spread to nearby lymph nodes.
- Stage III. The cancer has spread to nearby lymph nodes but has not yet affected other organs.
- Stage IV. The cancer has spread to other organs. This process of spread is called metastasis. The most common locations of metastases from colorectal cancer are the liver, the lungs, the inside of the abdomen, or the ovaries.

The next steps in treatment depend on the stage of the cancer. Most colorectal cancers are first treated by some type of surgery.

- Small Stage 0 cancers may be completely removed during a colonoscopy. Some larger polyps can also be removed by inserting surgical instruments through the abdominal wall in a procedure called a laparoscopy.
- Stage I or Stage II cancers may be treated by removing the section of the colon that contains the tumor and then reconnecting the cut ends of the bowel. If reconnection is not possible, or if the cancer is at the lower end of the rectum, the doctor may have to perform a

colostomy, in which an opening called a stoma is made in the wall of the abdomen and a portion of the remaining colon is attached to the stoma. The person's body wastes pass through the stoma and are collected in a special bag attached to the outside of the body.

- If the cancer is advanced, surgery is unlikely to cure it. However, the surgeon can remove some of the tumor in order to relieve pain and bleeding.
- If the colorectal cancer has spread only to the liver and the patient's health is otherwise good, the surgeon can remove the cancerous part of the liver with the colorectal tumor.

Radiation therapy or chemotherapy may be used following surgery to lower the risk of recurrence. Chemotherapy is often used to treat patients with Stage III or Stage IV cancer for a period of six to eight months after surgery. Radiation therapy is used more often to treat Stage III rectal cancer, although it may also be given to patients with colon cancer to relieve pain or to shrink tumors before surgery.

#### Prognosis

In spite of progress in early identification and treatment of colorectal cancer, it remains the third leading cause of death from cancer in the United States. Prognosis for recovery depends on the stage at which the disease is detected and treated. If it does not reappear (recur) within five years, it is considered cured. Stage I, II, and III colorectal cancers are considered potentially curable, but most doctors do not consider Stage IV cancer to be curable.

According to the NCI, 93 percent of colon cancer patients and 93 percent of rectal cancer patients who were diagnosed with Stage I cancer are still alive five years after diagnosis, but only 39 percent of colorectal cancers are detected at this early stage. The five-year survival rate drops to 8 percent for those diagnosed with Stage IV cancer.

#### Prevention

Screening tests for colon cancer in adults over age 50 are good preventive measures. The death rate for colon cancer has dropped since 1990, in part because of increased awareness and screening by colonoscopy. Colon cancer can almost always be caught in its earliest and most curable stages by colonoscopy.

# WORDS TO KNOW

Benign: Not cancerous.

**Colon:** The part of the large intestine that extends from the cecum to the rectum.

**Metastasis (plural, metastases):** The spread of cancer from its primary location to other parts of the body.

**Polyp:** A growth of tissue protruding from a mucous membrane such as the colon.

**Staging:** Measuring the severity or spread of a cancer.

**Stoma:** An opening made in the abdomen following surgery for colon cancer that allows wastes to pass from the body.

**Tumor markers:** Substances found in blood, urine, or body tissues that can be used to detect cancer.

People who have either of the two genes for early colorectal cancer, which can be identified by genetic testing, should have colonoscopies during their late teens and have their colons removed when they are in their 20s to prevent getting colorectal cancer.

Although further research is needed to find out how large a role diet plays in the development of colorectal cancer, people should consider lowering the amount of fat and increasing the amount of fiber in their diets to lower their risk of this type of cancer.

#### The Future

It is likely that the survival rate for patients with colorectal cancer will continue to rise over the next few decades. The introduction of more effective screening techniques, the development of new drugs to treat cancer, and further advances in surgical technique should all be beneficial.

SEE ALSO Breast cancer; Crohn disease; Gigantism

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# Common Cold

# Definition

The common cold is a contagious viral infection of the upper respiratory tract (nose and throat). It is self-limiting, meaning that it clears up by itself without the need for special medical treatment.

The disease got its name in the sixteenth century, when English doctors noticed that colds are more frequent in northern countries during the winter months and thought that exposure to low temperatures caused colds. It was not until the eighteenth century that Benjamin Franklin suggested that cold weather by itself does not cause colds but helps them to spread by driving people indoors where they are crowded more closely together. Although viruses had not been discovered by Franklin's day, he was correct in thinking that colds are transmitted from one person to another through the air or by direct contact.

# Description

The common cold is one of the most widespread infectious diseases in the world. It is caused by about 200 different viruses belonging to at least eight different families of viruses. For most people, the classic symptoms of a cold are an irritated nose or scratchy throat within eight hours to two days after infection, followed quickly by a runny nose and sneezing. Although many people experience headaches, general tiredness, and loss of appetite as well, the main symptoms of a cold are in the nose.

A person can get a cold by inhaling the virus directly if they are sitting close to an infected person who is sneezing or coughing. They can also get a cold by touching their eyes, nose, or mouth after touching an object or

#### Also Known As

Coryza, acute viral nasopharyngitis

#### Cause

Viruses

#### Symptoms

Sore throat, stuffed-up and runny nose, watery eyes, coughing, sneezing, low-grade fever

#### Duration

One to two weeks


*Computer model of the common cold virus.* AP IMAGES.

surface contaminated by the virus. Research indicates that cold viruses can live on skin for as long as two hours and on drinking glasses or other hard surfaces for as long as four days.

People with colds are most likely to spread the virus to others during the first two to three days of infection. After that they are much less contagious.

#### Demographics

No exact statistics are kept on the number of colds each year in the United States or in any other country because the illness is so common and many people take care of their symptoms at home. The Centers for Disease Control and Prevention (CDC) estimates that people in the

United States suffer about 1 billion colds every year; children lose between 22 and 189 million school days in an average year, and their parents lose 126 million work days to stay home and take care of them. Other employees miss an average of 150 million work days every year because of colds. The total impact of colds on the American economy comes to an estimated \$20 billion per year.

Colds are equally common in people of all races and ethnic groups. Some studies indicate that boys younger than three are more likely than girls to get colds in day care settings; however, in older children and adults, males and females are equally likely to get colds. In terms of age, children get colds more frequently than adults. Children average three to eight colds every year, and parents frequently get colds from their children. Colds become less frequent in later life, however; on average, people over sixty have less than one cold a year.

Colds are more common in North America during the fall and winter months, when children are in school and adults are spending more time indoors. In tropical climates, colds are most common during the rainy season, as humid conditions increase the viruses' survival time outside the human body.

#### **Causes and Symptoms**

The common cold is caused by at least 200 viruses that had been identified as of 2008. A cold virus typically enters the body through the nasal

## Home Care for Colds

Most people with colds do not need to see a doctor. They can relieve cold symptoms at home by using one or more of the following treatments:

- Rest at home rather than going to school or work. Trying to keep up a normal schedule of activities outside the house exposes other people to the cold virus.
- Drink plenty of fluids. Some people find herbal teas containing licorice, chamomile, lemon, or ginger soothing to inflamed throat tissues.
- Take a mild over-the counter pain reliever for headache, fever, or muscle cramps.
- Use a decongestant nasal spray to relieve a stuffed-up nose. These sprays should not be used for more than a few days, however, because they can dry out the membranes lining the nasal passages. In addition, they should not be used more frequently than recommended on the package. Overuse of

decongestants can lead to a rebound reaction, in which the nasal passages react to the decongestant by becoming even more swollen and irritated.

- Take an antihistamine. Some people are helped by such preparations as Benadryl or Nytol, which slow down the secretion of mucus and relieve a runny nose. Antihistamines do, however, make many people drowsy and are best used before bedtime.
- Cough medicines. These preparations should not be given to children because of the possibility of side effects but may provide temporary relief for adults from coughing due to a cold.

Although claims have been made that taking zinc, large amounts of vitamin C, or extracts of echinacea will help to cure a cold, there is no convincing evidence for the effectiveness of any of these substances.

passages or mouth, but can also enter through the mucous membranes covering the eyes. The virus spreads to the tissues lining the area between the nose and the back of the throat, where it rapidly multiplies. Within hours or a day or two after the virus enters the body, the infected person feels a scratchy sensation at the back of the throat, followed by sneezing and a constant flow of runny mucus from the nose. Researchers think that these symptoms are caused by the body's immune response to the virus rather than by tissue damage caused by the virus.

In addition to the runny nose, sneezing, and coughing associated with the common cold, people may also have:

- Low-grade fever (101°F [38.8°C] or lower)
- Muscle aches
- Headache
- Loss of the senses of taste and smell

- Loss of appetite
- Sore throat

Children are often sicker than adults when they get a cold because their immune systems are less developed. Children may run a fever as high as 102°F (38.9°C) with a cold; they may also develop an ear or sinus infection following a cold. Children with asthma may have an attack triggered by a cold.

#### Diagnosis

For most people, the diagnosis of a cold is obvious from its symptoms, particularly if they know they have recently been exposed to someone else with a cold. Most people do not need to see a doctor to be diagnosed with a cold. They should, however, see their doctor if they have any of the following symptoms, which may indicate an allergy or a more serious illness:

- The symptoms last longer than two weeks.
- The patient has a fever of 102°F (38.9°C) or higher.
- They are coughing up thick mucus.
- They have severely swollen glands.
- They have severe pain in the sinuses.
- They are having chills or night sweats, or are extremely fatigued.

#### Treatment

There is no cure for the common cold. Treatment is aimed at relieving the sneezing and other symptoms until the body's immune system clears the virus. The sidebar lists some common home and over-the-counter remedies that ease the symptoms of a cold.

Although many people ask their doctors for antibiotics to treat a cold, it is important to know that antibiotics are not effective against viruses. In fact, overprescribing of antibiotics is a major factor in the emergence and spread of antibiotic-resistant bacteria. The doctor will, however, prescribe antibiotics if the patient has developed a sinus infection caused by bacteria.

#### Prognosis

Most people recover from a cold in seven to eleven days with no longterm complications. Children, however, may develop earaches following a cold.

## Prevention

There is no vaccine effective against colds. The following precautions, however, can lower a person's risk of getting frequent colds:

- Stay away from people with colds whenever possible.
- Wash the hands frequently.
- Avoid touching the mouth and face after being exposed to someone with a cold.
- Use alcohol-based hand sanitizers.
- Keep kitchen and bathroom countertops and other surfaces clean.
- Wash children's toys after play.
- Do not share drinking glasses, cups, or food utensils. Use disposable paper cups when sick to protect other family members.

#### The Future

Several drug companies are working on antiviral drugs that might help people recover from colds more rapidly. One such drug is being tested in an oral form while a second drug is being developed that would be applied as a nasal spray. One limitation of these drugs, however, is that they would work only against cold viruses belonging to one of the eight groups known to cause the common cold.

The development of an effective vaccine against colds is considered unlikely. One reason is the sheer number of viruses known to cause colds. Another reason is that these viruses mutate (change their DNA) very rapidly; thus any vaccine that might be developed would be outdated by the time it entered clinical trials, let alone be approved for use.

SEE ALSO Asthma; Ear infection; Influenza; Sore throat

#### For more information

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## WORDS TO KNOW

**Echinacea:** A plant native to the eastern United States that is thought by some to be a useful cold remedy. It is also known as purple coneflower. **Mutate:** A change in the genetic material of an organism. Viruses can mutate rapidly.

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## Concussion

#### Definition

Concussion is defined as a closed-head injury (that is, one that does not involve a skull fracture) usually caused by blunt-force impact. Concussion was traditionally used by doctors to refer to temporary loss of consciousness from a head injury, but in everyday language, the term usually means any minor injury to the head or brain. Concussions generally are not life-threatening but may involve long-term as well as short-term health problems.

#### Description

Concussion occurs when the brain is pushed against one side of the skull, often by impact with a blunt object. The impact can result from a person's striking the head against an object as well as being hit by something or someone. Athletic injuries are the most common cause of concussions, but concussions can also result from automobile, bicycle, or other transportation accidents, criminal assaults, workplace accidents, or accidents in the home—particularly falls.

## **Demographics**

Concussions are probably underreported because many mild cases resolve on their own and because many athletes do not want to miss scheduled competitions. One estimate is that there are six cases of concussion per 1,000 people per year in the United States, but this figure is probably too low.

Among children between the ages of five and fourteen, the most common causes of concussions are sports and bicycle accidents; among adults, the most common causes are falls and auto accidents. Soldiers in combat may have rates of concussion from bomb blasts as high as 15 percent.

The sports that place participants at the highest risk of a concussion are boxing, football, ice hockey, wrestling, rugby, and soccer. At the high school level, girls are more likely to be injured than boys (in sports played by both sexes) because of their smaller head and neck structure. According to the American College of Sports Medicine, studies of high school athletes show that the rate of concussions per 1,000 games is as follows: 0.59 for football; 0.25 for wrestling; 0.18 for boys' soccer and 0.23 for girls' soccer; 0.09 for girls' field hockey; and 0.11 for boys' basketball and 0.16 for girls' basketball.

## **Causes and Symptoms**

The basic cause of a concussion is the rotation of the brain inside the skull as well as compression of the brain tissue against the sides of the skull when a person receives a blow on the head. Although the brain is cushioned against minor bumps by cerebrospinal fluid (CSF), the

## Also Known As

Mild traumatic brain injury, MTBI, minor head trauma

#### Cause

Blunt force trauma to the head

#### Symptoms

Headache, temporary loss of consciousness, ringing in the ears, confusion, nausea and vomiting, dizziness, visual disturbances

#### Duration

Several hours to several months

#### Concussion

WNBA player Chelsea Newton after suffering a mild concussion during a game. AP IMAGES.



amount of fluid between the brain and the skull is not enough to absorb the force of a heavy blow.

Doctors do not agree as to whether concussion is a disturbance of brain function only or whether it involves damage to the structure of the brain as well. It is presently thought that the forces from a blow to the head disrupt the normal activity of the cells in the brain tissue, and that it is this disruption that causes temporary alterations in consciousness.

Concussion

The brain's secretion of neurotransmitters (the chemicals that convey signals from one nerve cell to another), its oxygen supply, and its use of glucose (blood sugar) are all affected by concussion. In addition, a small number of cells in the brain tissue may die as a result of even temporary interruptions of blood flow to the parts of the brain affected by the concussion.

The symptoms of a concussion include emotional and well as cognitive and physical symptoms:

- Physical symptoms: Headache (the most common symptom of concussion), dizziness, loss of balance or coordination, nausea and vomiting, visual disturbances, ringing in the ears, seizures or convulsions.
- Cognitive: Temporary loss or alteration of consciousness, difficulty concentrating, memory loss, confusion, disorientation, difficulty thinking clearly, disturbed sleep patterns.
- Emotional: Moodiness, tearfulness, temper outbursts, emotions inappropriate to a specific situation (such as laughing at a funeral), long-term personality changes.

## Diagnosis

It is possible for a person to suffer a mild concussion without knowing it, particularly if their

attention was focused on something else—such as getting out of a damaged car, helping someone else to safety, or escaping from an attacker. They may not seek medical help afterward. Most people, however, will go to the doctor after a head injury if they have briefly lost consciousness, are having prolonged vomiting spells, severe headaches, or such vision problems as seeing double or seeing bright lights.

Because of the potential seriousness of a closed-head injury, the doctor will take a careful history of the injury (its cause and other details of the incident, as well as how long the patient was unconscious) as well

## **Concussions in Professional Sports**

Several deaths among high-profile professional athletes focused attention on the dangers of ignoring or minimizing concussions particularly the long-term effects of repeated head injuries. In 2006, Andre Waters, a professional football player, committed suicide in his Florida home. Waters's suicide was preceded in 2004 by the death of Justin Strzelczyk, another football star, in an auto accident following a 37-mile-long high-speed police chase on the wrong side of the highway.

Christopher Nowinski, a former professional wrestler whose career was ended by post-concussion syndrome in 2003, decided to investigate the effects of repeated concussions on professional athletes. Nowinski contacted a neurosurgeon at the University of Pittsburgh who was able to obtain samples of the dead football players' brain tissues after contacting their families. The doctor found that Waters's brain resembled that of an 85-year-old man with Alzheimer disease, and that Strzelczyk's personality changes in the weeks preceding his death could be traced to brain damage from repeated concussions. In 2007 Nowinski founded the Sports Legacy Institute, an organization dedicated to researching sports-related brain injuries in order to protect the health and lives of all athletes.

as a general medical history. The doctor will particularly need to know whether the patient has had previous head injuries; is taking any bloodthinning medications; or has a bleeding disorder. The history is followed by several examinations and imaging studies:

- General physical examination. The doctor looks for injuries to the head and neck, including bleeding from the ears or nose, or other indications that the skull may have been fractured.
- Neurological examination. This part of the examination involves testing the patient's reflexes, vision, hearing, balance, and coordination.
- A computed tomography (CT) scan. This type of imaging study is not always ordered if the concussion seems to have been mild. It is, however, usually performed if the patient is a child or elderly; is under the influence of alcohol or drugs; was injured in a motor vehicle accident; fell from a height of more than 3 feet (0.9 meters); cannot recall the accident or blow thirty minutes after it occurred; is having seizures; is vomiting; or is having trouble with short-term memory.
- Observation overnight in the hospital or at home for twenty-four to forty-eight hours. This is done to make sure that the patient does not have a more severe injury than was first thought. The doctor may ask a friend or relative of a patient sent home to wake them every few hours for the first thirty-six hours after the injury to make sure that they can return to full consciousness.
- Magnetic resonance imaging (MRI). This type of imaging study may be performed if the patient's headaches and other symptoms are getting worse or have lasted a week or longer.

#### Treatment

Treatment of mild concussion usually consists of a mild over-the-counter pain reliever such as acetaminophen (Tylenol) or ibuprofen (Advil) plus bed rest and extra fluids. Aspirin should be avoided because it increases the risk of bleeding. If the patient has a bruise under the skin of the head (a "goose egg"), ice wrapped in a washcloth can be applied to relieve the swelling. Cuts in the skin may be treated in the doctor's office with a local anesthetic and stitches if necessary.

People injured in an athletic competition should not play again for at least seven to ten days after the concussion.

## Prognosis

Prognosis depends on the severity of the concussion. Most people heal completely in one to two weeks, although healing may be somewhat slower in adults over fifty-five. About 10 percent of patients develop what is known as post-concussion syndrome (PCS). This is a condition marked by persistent headaches for two weeks to several months after the concussion. The patient may also have nausea and vomiting, or difficulty reading or concentrating on their work. PCS usually goes away on its own. If it persists after a few months, the patient will usually be referred to a neurologist for further evaluation.

The effects of concussions tend to add up over time; it is known, for example, that each concussion a person sustains makes it easier for them to suffer another concussion in the future. Repeated concussions can lead to permanent loss of memory and depression or other psychiatric disorders. Professional athletes are particularly likely to suffer long-term injuries from repeated concussions; several studies have found that the rate of depression is three times higher and the risk of Alzheimer disease five times higher in athletes who have had three or more concussions.

## Prevention

Not all accidents or other causes of concussions can be prevented. People can, however, lower their risk of concussions by taking the following safety measures:

- Sports: Athletes should wear protective headgear for such sports as martial arts, football, baseball, bicycling, motorcycling, rollerblading, and skateboarding. They should make sure their helmets and other protective gear are properly fitted. Coaches should insist that players are taught the rules of safe play and that they follow them. Last, players should report all head injuries to the team doctor even if they seem minor at the time.
- Workplace: People in occupations requiring hardhats should wear them at all times, particularly in construction work. Even a small object dropped from the upper floors of a building can cause a serious head injury.
- Home safety: Elderly persons or parents of small children should check their house or apartment for loose rugs, poor lighting, slippery floors, and other problems that increase the risk of falls—which are a common cause of concussions.

### WORDS TO KNOW

**Closed-head injury:** A head injury that does not involve fracturing of the skull.

**Cognitive:** Related to thinking, memory, and other conscious intellectual activities or processes.

**Post-concussion syndrome (PCS):** A condition characterized by several weeks or months of headache following a head injury.

#### The Future

Concussions are likely to continue to be relatively commonplace head injuries, particularly in victims of bicycle, motorcycle, and auto accidents. Stricter regulations about allowing injured athletes to play, however, may reduce the rate of concussions and their long-term consequences in amateur as well as professional sports.

SEE ALSO Alzheimer disease; Depression; Shaken baby syndrome

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# U

## **Congenital Heart Disease**

## Definition

Congenital heart disease, also known as congenital heart defect or CHD, refers to problems or abnormalities in the structure of a baby's heart that are present at birth. These abnormalities can affect the heart muscle itself, the valves inside the heart, or the arteries and veins supplying the heart. The defects can affect the baby's health by slowing down the flow of blood, blocking the blood flow completely, or causing the blood to flow in the wrong direction. Congenital heart defects are the most common type of major birth defect; they range from minor problems that do not cause any obvious symptoms to life-threatening abnormalities.

Doctors divide congenital heart defects into two major categories depending on whether they cause a bluish-gray discoloration of the mouth, lips, and nail beds called cyanosis. Cyanosis occurs when the blood flowing through the blood vessels close to the skin surface is not carrying enough oxygen to give the lips or mouth their normal pinkish color. Some congenital heart defects lead to cyanosis while others are called noncyanotic defects because they do not produce this skin discoloration.

## Description

The signs and symptoms of congenital heart defects vary greatly in severity. Most of these defects produce a heart murmur—a characteristic whispering sound that the doctor can hear through a stethoscope. Some defects, however, may produce no other noticeable symptoms, while others may lead to shortness of breath, cyanosis, chest pain, sweating, frequent respiratory infections, underdeveloped limbs and muscles, poor feeding, or slowed growth.

## Also Known As

Congenital heart defect, CHD

#### Cause

Unknown for most; genetic abnormalities, mother's infection, or drug abuse in some cases

#### Symptoms

Bluish lips and tongue, weak pulse, difficulty feeding or breathing, heart murmur

#### Duration

Some are lifelong unless treated, others improve without treatment

#### **Congenital Heart Disease**

Image showing a congenital heart defect, in this case a hole in the septum, which separates the left and right sides of the heart. © SIMON FRASER/SPL/ PHOTO RESEARCHERS, INC.



Cyanotic congenital heart defects include:

- Tetralogy of Fallot. This is the most common cyanotic heart defect, accounting for 5–7 percent of all congenital heart defects. It is named for a French doctor who described it in 1888. Tetralogy means that this defect involves four different interrelated abnormalities: a hole between the two bottom chambers (ventricles) of the heart; an aorta in an abnormal location; a narrowing of a heart valve that prevents normal blood outflow from the right ventricle; and a larger than normal right ventricle.
- Transposition of the great vessels (TGA). TGA is a defect in which the aorta and the pulmonary artery (the artery that carries blood from the heart to the lungs) are switched, so that the heart receives blood that has not been oxygenated in the lungs and sends this oxygen-poor blood out to the rest of the body.

- Persistent truncus arteriosus. The truncus arteriosus is a structure found in human embryos that normally divides to form the aorta and the pulmonary artery prior to birth. If that division does not occur, the baby is born with a single large blood vessel instead of two normal arteries and can develop heart failure within a few weeks after birth.
- Tricuspid atresia. Tricuspid atresia is a heart defect in which the valve between the two chambers on the right side of the heart does not open. In addition, the heart's right ventricle is unusually small.

Noncyanotic congenital heart defects include:

- Ventricular septal defect (VSD). A VSD is a hole in the wall of tissue that normally divides the two lower chambers of the heart. It is the most common type of CHD, accounting for 30–60 percent of all CHDs.
- Atrial septal defect (ASD). An ASD is a hole in the tissue that normally separates the two upper chambers of the heart.
- Patent ductus arteriosus (PDA). The ductus arteriosus is a temporary passageway that connects the pulmonary artery and the aorta that usually closes within a few hours or days after the baby is born. Patent means that the passageway remains open. PDAs are more common in babies born prematurely than in full-term babies.
- Aortic stenosis. Aortic stenosis is a defect in which the aortic valve—a heart valve that controls the flow of blood from the left ventricle to the aorta—is narrower than normal and slows down the flow of blood out of the heart.
- Coarctation of the aorta (COA). In COA, a portion of the aorta is abnormally narrow, reducing the amount of blood that the heart can pump to the rest of the body.

## **Demographics**

Congenital heart disease is one of the most common birth defects, occurring in eight out of every 1,000 newborns in the United States, or about 40,000 babies in an average year according to the March of Dimes. Most congenital heart defects either heal by themselves or respond well to treatment; however, 4,000 babies with CHDs will not survive their first year of life. Twice as many children die each year from CHDs than from all types of children's cancers combined. Risk factors for congenital heart disease include:

- Premature birth.
- Family history of CHD. Having one baby with a CHD increases the risk of having a second child with a heart defect by a factor of five. If the baby's mother has a CHD, the risk for a child to be born with CHD ranges 2.5–18 percent, with an average risk of 6.7 percent. If the father has CHD, the risk for a child to be born with CHD ranges 1.5–3 percent.
- Rubella in the mother during early pregnancy.
- Mother's abuse of alcohol or drugs during pregnancy.
- Chronic diseases in the mother, particularly diabetes and lupus.
- Mother's use of certain antiseizure medications.

## **Causes and Symptoms**

Most cases of congenital heart disease (85–90 percent) do not have a known cause. Some known causes of CHDs include:

- Genetic disorders involving parts of a chromosome (5–8 percent of CHDs): these include Down syndrome, Edwards syndrome, Patau syndrome, and Turner syndrome.
- Single-gene defects (3–5 percent): Marfan syndrome is the most common single-gene disorder linked to CHDs.
- Environmental factors (2 percent).

Symptoms of CHDs vary somewhat depending on the specific heart defect but may include:

- Heart murmur
- Cyanosis
- Rapid breathing
- Lack of appetite and poor feeding
- Failure to gain weight
- Sweating during feedings
- Weak pulse

#### Diagnosis

Some CHDs can be detected during pregnancy or during the baby's first physical examination when the doctor listens to the heart and looks for cyanosis, rapid breathing, a weak pulse, or indications of heart failure. Small heart defects may not be detected until the child is older or even until the adult years.

Specific tests that can be done to evaluate the type and severity of a congenital heart defect include:

- Echocardiogram. This is a test that uses sound waves to produce an image of the baby's heart. It can be done during the fourth or fifth month of pregnancy to plan treatment for the baby after it is born. An echocardiogram can also be performed to diagnose babies after birth.
- Chest x ray. This test may be done to see whether the baby's lungs are normal as well as whether its heart is abnormally large.
- Electrocardiogram (ECG or EKG). An ECG is a tracing of the heart's electrical activity. It shows the speed of the heartbeat and whether the heart rhythm is regular or irregular.
- Pulse oximetry. This test uses a sensor placed on the baby's fingertip or toe to measure how much oxygen is carried in the blood.
- Cardiac catheterization. A catheter is a long thin tube that is passed through a vein in the baby's neck, arm, or upper thigh to reach the heart. A special dye that can be seen on x ray is injected through the catheter into the blood vessels or the heart itself. This test allows the doctor to trace the path and the direction that the blood takes through the chambers and major blood vessels of the heart.

#### Treatment

Treatment of congenital heart disease varies depending on the type of defect and its severity. Some small defects may not require any specific treatment. In some cases the doctor may prescribe medications—usually diuretics to help the baby eliminate excess water through the urine; or digoxin, a drug that helps the heart muscle contract with greater force, thus strengthening the heart.

Some congenital heart defects require one or more surgical procedures to repair the abnormalities. Atrial septal defects and aortic valve stenosis can be repaired by using a catheter. The surgeon inserts the catheter through a vein and threads it into the heart. A small device can be positioned through the catheter to fill an ASD, or a balloon can be threaded through the catheter to open a partially closed valve. The balloon can then be collapsed and the catheter removed.

Open-heart surgery is usually necessary to replace a badly damaged valve; to close large holes in the septa between the ventricles or atria; or to repair a complicated defect like tetralogy of Fallot. In a few very rare cases, the baby may need a heart transplant if there are multiple defects that are too severe or complicated to repair by surgery.

#### Prognosis

The prognosis of congenital heart disease varies. About 10 percent of infants with CHDs die during their first year of life. Advances in surgery since the 1980s mean that most children with congenital heart disease can live into adulthood and enjoy normal and productive lives. Many do not need any special care, although some will need periodic checkups with a heart specialist as well as with their pediatrician as they grow older.

## Prevention

Most CHDs cannot be prevented because their cause is still unknown. Some can be prevented, however, by the following measures:

- Vaccination against rubella before getting pregnant.
- Avoiding alcohol and drugs during pregnancy.
- Consulting a genetic counselor before becoming pregnant if there is a family history of CHD.
- Consulting a doctor before becoming pregnant if the mother is being treated for diabetes, lupus, or seizure disorder.

## The Future

Congenital heart disease is not likely to become more common in the general population in the near future. Research in the field is directed toward a better understanding of the genetic factors that may be involved in CHD, and closer study of the development of a baby's heart during pregnancy. This information in turn may guide the invention of new or improved surgical techniques to treat congenital heart defects as early as possible, perhaps even before the baby's birth.

## WORDS TO KNOW

- **Aorta:** The large artery that carries blood away from the heart to be distributed to the rest of the body.
- Atrium (plural, atria): One of the two upper chambers of the heart.

Congenital: Present at birth.

**Cyanosis:** A blue discoloration of the lips, inside of the mouth, and nail beds caused by lack of oxygen in the blood vessels near the skin surface.

- **Embryo:** The medical term for an unborn baby from the time of conception to the end of its first eight weeks of life.
- Patent: The medical term for open or unobstructed.
- **Septum (plural, septa):** A partition that separates two cavities or chambers in the body.
- **Stenosis:** The medical term for abnormal narrowing of the opening of a blood vessel.
- **Ventricle:** One of the two lower chambers of the heart.

**SEE ALSO** Down syndrome; Edwards syndrome; Fetal alcohol syndrome; Marfan syndrome; Patau syndrome; Prematurity; Rubella; Seizure disorder; Turner syndrome

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# U

# **Conjoined Twins**

## Definition

Conjoined twins are identical twins in which the two embryos fail to separate completely before birth. It is thought that the condition results from incomplete splitting after the twelfth day of fetal development. If a fertilized human egg divides into two embryos before the twelfth day, the identical twins will be born normally as two separate infants. The longer the delay in the separation of the two embryos, the more complicated the connections between the conjoined twins are likely to be.

Conjoined twins can be categorized in several different ways. One distinction that is often made is between symmetrical (equal) conjoined twins, in which both infants are well developed; and asymmetrical (unequal) conjoined twins, in which an incomplete twin is joined to a fully developed sibling. Conjoined twins are also classified according to the points at which their bodies are joined. This system goes back to a French biologist named Étienne Geoffroy Saint-Hillaire (1772–1844), who based his terms on the formal medical words for parts of the body. These categories are listed in the next section.

## Description

Conjoined twins may share tissue, organ systems, or both, depending on the points at which their bodies are fused:

- Thoracopagus: 40 percent of conjoined twins are joined at the chest. These twins always share a heart.
- Omphalopagus: This term refers to twins joined at the abdomen, and these account for 34 percent of cases. These twins often share a liver and digestive system.
- Pygopagus: The twins are joined back to back at the buttocks and account for 18 percent of cases. These twins sometimes share the lower part of the digestive tract or the genital organs.
- Ischiopagus: The twins are joined at the pelvis or lower spine with the lower half of the bodies fused. They have four arms but may have between two and four legs. Six percent of cases fall into this group.

#### Also Known As

Siamese twins (now considered culturally inappropriate)

#### Cause

Delayed splitting of a fertilized human egg

#### Symptoms

Two bodies joined at various locations

#### Duration

Lifelong unless corrected by surgery

#### **Conjoined Twins**

<image>

Iranian conjoined twins Laden and Laleh Bijani. AP IMAGES.

- Craniopagus: Two percent of conjoined twins are fused at the head but the bodies are separate. In some cases these twins share part of the brain as well as the skull.
- Parasitic twins: This term is used to describe cases in which one twin is much smaller than the other and depends on the larger twin for survival. Some researchers estimate that 10 percent of conjoined twins fall into this category.

#### **Demographics**

Estimates vary somewhat, but most researchers think that conjoined twins occur once in every 33,000–165,000 births; however, 40–60 percent of conjoined twins are stillbirths. In the United States, one in every 200,000 live births is a set of conjoined twins.

Male conjoined twins are more likely to be stillbirths. Conjoined twins that are born alive are three times more likely to be females.

Conjoined twins are more common in India and Africa than in Europe or North America. The reasons for this difference are not known.

## Chang and Eng

Chang and Eng Bunker (1811–1874) were conjoined twins born in 1811 in Thailand (formerly called Siam), for whom the term "Siamese twins," a term that is no longer culturally accepted, was coined. Joined at the breastbone by a band of cartilage, their vital organs functioned independently; they could have been easily separated if modern surgical techniques and equipment had been available.

Like some other nineteenth-century conjoined twins, Chang and Eng were discovered by a British business promoter in 1829 and exhibited as a medical curiosity. They later joined P.T. Barnum's traveling circus. They left the circus in 1839, settling in North Carolina. Taking the surname Bunker, they became U.S. citizens and married two sisters in 1843. The wives did not get along, so the brothers set up separate households, spending three days each week in each home. Chang had ten children by his wife; Eng had eleven.

The twins' health declined in the early 1870s. Chang died on January 17, 1874; Eng died an hour later. They are subjects of several literary works, the first was a short story by Mark Twain



*Chang and Eng Bunker, Siamese twins.* BLANK ARCHIVES/ GETTY IMAGES.

called "The Siamese Twins." With characteristic humor, Twain suggested that the twins had fought on opposite sides during the Civil War, and that one was fifty-three years old while his twin was only fifty-one.

#### **Causes and Symptoms**

Identical twins develop when a fertilized human egg splits into two separate embryos during the first twelve days following conception. If the split occurs after the twelfth day, the twins will not separate fully from each other and will be conjoined at birth. The cause of most cases of delayed splitting is not known; no genes have yet been identified that lead to conjoined twins. There have been, however, about ten cases reported in which conjoined twinning occurred after the mother had been exposed to a drug given to stimulate ovulation or a drug used to treat fungal infections.

The symptoms of conjoined twins depend on the location of the fusion between the two twins and whether the twins are symmetrical (equal) or not. In many cases the twins die shortly after birth from heart failure, lung failure, or obstruction in the digestive tract.

#### Diagnosis

In some cases the doctor may suspect the presence of conjoined twins early in the pregnancy by finding that the mother's uterus is larger than expected and hearing two fetal heartbeats when listening through a stethoscope. Conjoined twins can be diagnosed by ultrasound as early as the eighth week of pregnancy. Magnetic resonance imaging (MRI) can be used to identify which organ systems are shared between the twins. Computed tomography (CT) scans are primarily useful in evaluating the bony structures of twins fused at the hips or pelvis.

Electrocardiograms (ECGs) and electroencephalograms (EEGs) may be used to evaluate the extent of shared heart or brain function and to determine whether surgical separation of the twins is possible.

#### Treatment

The treatment of conjoined twins is highly individualized. It almost always requires complicated surgery in one of a small number of medical centers (three in the United States, one in the United Kingdom, one in New Zealand, and one in South Africa). Doctors classify conjoined twins for treatment in one of three categories: those who will die shortly after birth; those who require immediate emergency surgery; and those who will survive until they are old enough for surgery to have a higher chance of success (usually six to twelve months). Twins who share a heart or brain usually cannot be separated without causing the death of both twins.

Conjoined twins are usually delivered by cesarean section rather than waiting for the mother's due date. The operation is usually scheduled for two to four weeks before the due date. Surgery to separate the twins may be performed immediately after delivery if one or both twins have a life-threatening emergency. These operations are complicated and may take as long as thirty-five hours to complete. Two complete surgical teams are required to care for the twins after the separation is complete. In most cases, conjoined twins who survive separation will need further surgery at intervals during childhood.

#### Prognosis

The prognosis of conjoined twins is often poor. Between 40 and 80 percent of twins who need emergency surgery after birth die in intensive care following the operation. In some cases, particularly those involving parasitic twins, the parents must make the painful decision to allow one of the twins to die if the other is to have any chance of survival. Conjoined twins who are healthy enough to have separation surgery postponed until they are older have a survival rate of 80 percent.

It is possible for conjoined twins who are not separated to have productive and satisfying lives. One set of conjoined twins in Minnesota completed high school in 2008 and obtained a driver's license. One twin in a set of conjoined sisters has made a career as a country music singer. Conjoined twins who are not separated, however, have shortened life expectancies; most pairs die in their twenties or early thirties. As of 2008, the oldest known living set of conjoined twins was a pair of brothers in Ohio born in 1951.

#### Prevention

There is no known way to prevent conjoined twins other than avoidance of the small number of drugs that have been associated with conjoined twinning in a few cases. The condition is considered to be sporadic (occurring at random); parents do not have an increased risk of having a second set of conjoined twins in a later pregnancy.

#### The Future

The widespread use of ultrasound and other imaging techniques has led to improved prenatal identification and diagnosis of conjoined twins. It is possible that the greater numbers of conjoined twins that are identified before birth will help researchers understand the causes of this type of twinning and discover better methods of surgical separation and treatment for conjoined twins.

SEE ALSO Spina bifida

## WORDS TO KNOW

**Identical twins:** Twins that develop from a single fertilized egg that divides to form two separate embryos.

Sporadic: Occurring at random.

**Stillbirth:** The birth of a baby that has died before or during delivery.

Prenatal: Before birth.

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# **m** Conjunctivitis

## Definition

Conjunctivitis is an infection or inflammation of the conjunctivae, the thin clear membranes that cover the white part of the eyeballs and line the inside of the eyelids.

## Description

Conjunctivitis is a medical condition in which the conjunctivae appear pink or red, and the eyes water and feel irritated. In some cases the eyes may feel scratchy or itchy as well. Conjunctivitis can be caused by a bacterial, chlamydial, or viral infection; by seasonal allergies; by a foreign object in the eye; by a chemical irritant; or by such other medical conditions as dry eye. There may be mild sensitivity to light, a condition known as photophobia.

## Demographics

There are no exact statistics on the number of cases of conjunctivitis in the United States each year because the condition has so many possible causes, but it is the single most common reason for a person's consulting an eye doctor. Between 1 and 2 percent of babies born in the United States each year have ophthalmia neonatorum, a bacterial conjunctivitis caused by a sexually transmitted disease in the mother.

## **Causes and Symptoms**

Conjunctivitis may have a number of different causes. Some are not contagious, while others can be transmitted from person to person. The symptoms also vary somewhat depending on the cause:

- Allergic conjunctivitis. Allergic conjunctivitis is caused by pollen, dust, and the other types of allergens that cause hay fever and other seasonal allergies in some people. Both eyes are usually affected. The eyes are usually very watery and itchy in this type of conjunctivitis, and the eyelids may swell up or look puffy.
- Viral conjunctivitis. This type of conjunctivitis is contagious and often gets started when viruses from an upper respiratory infection

Also Known As Pink eye, red eye

#### Cause

Viruses, bacteria, allergies, chemicals or foreign objects in the eye, other eye disorders

#### Symptoms

Redness, itchiness, blurred vision, scratchy feeling in one or both eyes

#### Duration

A few days to a few weeks, depending on the cause

#### Conjunctivitis



*An eye with severe conjunctivitis.* © MEDICAL-ON-LINE / ALAMY.

like a cold are carried into the eye by hand-to-eye contact. The infection usually starts in one eye but often spreads to the other within a day or two. The discharge from the eye is usually watery. A person with viral conjunctivitis is contagious for one to two weeks after their symptoms first appear.

- Bacterial conjunctivitis. Like viral conjunctivitis, this type is also contagious and can be spread by sexual contact with someone infected with gonorrhea or chlamydia as well as by hand-to-eye transmission. Pregnant women infected with gonorrhea or chlamydia can give their babies a particularly dangerous type of bacterial conjunctivitis during childbirth known as ophthalmia neonatorum, which can cause blindness if untreated. Bacterial conjunctivitis is often accompanied by a gritty or sandy feeling in the eyes and a yellowish or grayish pus-filled discharge that causes the eyelids to stick together during sleep. Bacterial conjunctivitis usually appears within three days after the person is infected with the bacteria. Like viral conjunctivitis, it often begins in one eye but can spread to the other.
- Chemical splash (toxic conjunctivitis). This type of conjunctivitis is noninfectious. It is caused by getting an irritating chemical in the eye (such as shampoo or chlorine from swimming

## **Tips to Prevent Conjunctivitis**

To lower the risk of getting conjunctivitis or spreading it to others, eye doctors recommend the following practices:

- Change pillowcases frequently and do not share bed pillows with others.
- Do not share eye cosmetics, cotton balls, gauze pads, or facial tissues.
- Replace mascara, eye liner, and eye shadow regularly even if the cosmetics have not been completely used up.
- Do not share towels, handkerchiefs, or washcloths.
- People who wear contact lenses should handle and clean them properly.
- Keep hands away from the eyes as much as possible.
- Wash the hands regularly.

in pool water) and usually affects only the lower part of the conjunctiva inside the lower eyelid. Redness of the affected eye is sometimes caused by flushing the eye to wash out the chemical.

- Foreign body in the eye. Like toxic conjunctivitis, this type of conjunctivitis is not infectious. It usually affects only one eye and may be accompanied by a mucus-like discharge.
- Chronic or recurrent conjunctivitis. Some people develop recurrent episodes of conjunctivitis from wearing contact lenses or overusing certain types of eye drops. This type of conjunctivitis is not contagious.
- Conjunctivitis associated with dry eyes or other eye disorders. Some eye disorders are associated with a gradual loss of normal tear secretions in the eye, which can lead to conjunctivitis.

#### Diagnosis

In many cases conjunctivitis will clear up by itself in three to four days without the need to see a doctor. If the condition does not clear by itself, or if the discharge from the eye contains pus rather than being clear and watery, the person should see their doctor.

The doctor may take a sexual history or ask about recent exposure to colds and other viral illnesses or the patient's use of eye cosmetics or contact lenses to narrow the diagnostic possibilities. The doctor may also take a sample of the discharge from the affected eye or eyes for laboratory analysis in order to determine whether a bacterium or a virus is the cause.

Severe pain, intense photophobia, and blurred vision are not usually present in uncomplicated conjunctivitis. A patient with these symptoms should be examined further for more serious eye disorders, including glaucoma or inflammation of the interior of the eye. If the patient's eye has been irritated by a chemical, the doctor may examine the eye with a slit lamp to see whether the interior of the eye has also been affected.

#### Treatment

Treatment of conjunctivitis depends on the cause of the infection or irritation. Allergic or seasonal conjunctivitis is usually relieved by applying cool compresses to the eyes or by using artificial tears. This type of conjunctivitis can also be treated with antihistamines, steroids, or other antiinflammatory medications.

Bacterial infections are treated with antibiotic eye ointments or drops. There are no specific treatments for viral conjunctivitis; the illness must simply be allowed to go away on its own. Warm compresses or artificial tears may help to ease the patient's discomfort. Viral conjunctivitis may get worse for the first three to five days of the infection and then gradually get better; it can take as long as three to four weeks for viral conjunctivitis to clear up completely.

Foreign bodies or chemicals in the eye are usually washed out with saline solution. A chemical splash involving a caustic substance like lye is a medical emergency and should be treated by an emergency room physician or eye specialist as soon as possible.

#### Prognosis

The prognosis of conjunctivitis depends on its cause. Most people recover with no problems provided they follow the doctor's instructions for their particular type of conjunctivitis and practice good eye hygiene in general. The only type of conjunctivitis in adults that may cause permanent loss of vision is a chemical splash injury involving caustic chemicals such as lye or potash.

Ophthalmia neonatorum, however, can lead to blindness if it is not treated promptly. The usual treatments for bacterial infections of the eyes in newborns are silver nitrate eye drops or antibiotic medications.

#### Prevention

While it is not always possible to prevent being exposed to people with upper respiratory infections in one's school or workplace, one can lower one's risk of conjunctivitis by keeping the hands clean, being careful when touching the eyes, following instructions for the proper use and cleansing of contact lenses, and avoiding sharing washcloths and other personal items with others. Additional guidelines for preventing conjunctivitis are listed in the sidebar.

#### WORDS TO KNOW

- **Allergen:** Any substance that causes an allergic reaction in a person or animal.
- **Chlamydia:** A sexually transmitted disease caused by a bacterium that is a common cause of eye infections.
- **Conjunctiva (plural, conjunctivae):** The clear membrane that covers the white part of the eyeball and lines the eyelids.
- **Ophthalmia neonatorum:** The medical name for bacterial conjunctivitis in newborn babies caused by a sexually transmitted infection in the mother.
- **Photophobia:** A feeling of discomfort or pain in the eyes during exposure to light.
- **Slit lamp:** An instrument that focuses light into a thin slit. It is used by eye doctors to examine eyes for a wide variety of disorders.

#### The Future

Conjunctivitis has so many different causes that it is likely to continue to be a commonplace health concern for the foreseeable future.

SEE ALSO Chlamydia; Glaucoma; Gonorrhea; Hay fever

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## COPD

#### See Chronic obstructive pulmonary disease.

# m

## Coronary Artery Disease

## Definition

Coronary artery disease, or CAD, is a condition in which a fatty substance called plaque builds up inside the walls of the arteries that supply the heart. The plaque begins to form when the inner layer of tissue in the artery is damaged by smoking, diabetes, high blood pressure, personality factors, or some other cause. When the deposit of plaque grows large enough to narrow or block the flow of blood through the artery, the heart muscle becomes starved for oxygen, producing angina, difficulty breathing, or a full-blown heart attack.

#### Description

The buildup of plaque that leads to coronary artery disease can start as early as childhood. Arteries in the human body are blood vessels that carry blood away from the heart, in contrast to veins, which carry blood toward the heart. Arteries have three layers of tissue: an outer layer made of connective tissue, a middle layer made of smooth muscle, and a thin inner layer of cells that serve as a lining to the artery. This inner layer smoothes the flow of blood, which allows the heart to pump the blood further. The coronary arteries branch off from the base of the aorta, the large trunk artery at the top of the heart. They are the only source of blood supply to the heart muscle itself, which is why a blockage in these arteries is such a critical situation.

If the thin inner layer of a coronary artery is damaged, the body tries to heal it by covering it with a layer of plaque. Over time, the plaque deposit can become thicker, to the point where blood no longer flows smoothly through that part of the artery and the heart muscle becomes ischemic. Ischemia is a condition in which a portion of heart muscle (or any other tissue) is not receiving enough blood because of the blockage of an artery that ordinarily supplies it with blood. Ischemia in the heart can lead to angina—a type of chest pain that feels like squeezing or pressure and can move from the chest area to the arms, neck, jaw, or back.

The plaque deposits within the damaged artery can also rupture or crack open. Blood cells called platelets move to the ruptured area and form blood clots that block the artery. The loss of the blood supply following the closure of the artery leads to the death of heart tissue. The Also Known As CAD, coronary heart disease

#### Cause

Damage to the inner tissue layer of one of the coronary arteries

#### Symptoms

Angina (chest pain), shortness of breath, heart attack

Duration Years



death of part of the heart muscle is called a heart attack, which doctors call a myocardial infarction or MI.

## **Demographics**

CAD is the leading cause of death in the United States for both men and women. According to the American Heart Association, nearly fifteen million adults have some form of coronary artery disease. CAD usually develops in adults over thirty, and is common in adults over sixty. Coronary artery disease accounts for 650,000 deaths in the United States each year—25 percent of deaths among adults over the age of thirty-five.

Risk factors for coronary artery disease include:

- Age. The risk of coronary artery disease rises for men after age forty-five and for women after age fifty-five.
- Sex. Men are at greater risk of CAD throughout life; however, the risk for women rises after menopause.
- Family history of CAD. A person's risk of CAD is higher if their father or brother was diagnosed with CAD before age fifty-five, or a mother or sister diagnosed before age sixty-five.
- High blood pressure and high blood cholesterol levels.
- Race. African Americans, particularly African American women, and Native Americans have higher rates of CAD than either Caucasians or Hispanics.

- Lifestyle. People who smoke and drink heavily are at increased risk of CAD. Women who smoke a pack of cigarettes per day are six times as likely to have a heart attack as nonsmoking women; male smokers are three times as likely to develop CAD as nonsmokers.
- Obesity.
- Diabetes.
- Sleep apnea.
- Physical inactivity.
- High levels of emotional stress.
- Personality. People who are aggressive, impatient, and highly competitive are more likely to develop CAD than those who are relatively calm and relaxed.

#### **Causes and Symptoms**

Coronary artery disease is caused by a slow buildup—over years or decades—of plaque along the inner wall of a coronary artery. The plaque can block the flow of blood through the artery either by becoming thick enough to narrow the artery, or by rupturing and leading to the formation of a blood clot that blocks the artery.

Coronary artery disease can progress (get worse) for years without producing any noticeable symptoms. When symptoms do appear they typically take three forms:

- Angina or chest pain. Doctors distinguish two types of angina: stable angina, which occurs during exercise, after a heavy meal, or at other predictable times; and unstable angina, which varies in severity, timing, or frequency. Unstable angina is often an early warning of a heart attack, and requires emergency medical evaluation and treatment.
- Shortness of breath.
- A heart attack.

Some patients with CAD also experience lightheadedness or dizziness, an irregular heartbeat, or a racing heartbeat.

#### Diagnosis

For some people, a heart attack is the first symptom of CAD. They will be taken to a hospital emergency room, where they will be asked to describe their symptoms. In addition to taking the patient's personal and family history of risk factors for CAD, the doctor will also take the patient's temperature, blood pressure, and pulse. Listening to the patient's lungs and heartbeat through a stethoscope can help to rule out pneumonia or other diseases that might cause chest pain or difficulty breathing.

The next step is diagnostic tests to rule out a heart attack, which include:

- Electrocardiogram (ECG or EKG). An ECG or EKG measures the heart's electrical activity. Injured heart muscle makes unusual patterns or tracings on the paper printout produced by the ECG machine. If only a small amount of the heart muscle has been affected, however, the ECG may not show any abnormal patterns.
- Blood tests. These are done to confirm the diagnosis of a heart attack or to make sure that the electrocardiogram did not miss a small heart attack. Injured heart muscle leaks small amounts of special enzymes into the bloodstream.
- Chest x ray. A chest x ray may be done to see whether the patient's lungs are normal.
- Coronary angiography. Coronary angiography is a type of x-ray study in which the doctor threads a long thin tube called a catheter into the heart through an artery in the arm or upper thigh. A dye that will show up on x ray is injected into the blood-stream through the catheter. This test allows the doctor to find the location of the blockage in the coronary artery. It is also the only test that allows a heart specialist to determine the best treatment for the blockage.

If the patient is having angina but does not appear to be having a heart attack, he or she will be asked to take a stress test. In a stress test, the patient walks on a treadmill or pedals a stationary bicycle while hooked up to an ECG machine, which measures the electrical tracings of the heart before, during, and after exercise. Another type of stress test is a radionuclide stress test, in which a radioactive tracer element is injected into a vein while a special camera records the amount of the trace element that reaches various parts of the heart.

## Treatment

CAD can be treated with a combination of lifestyle changes, medications, and possibly surgery. Lifestyle changes include quitting smoking, losing

weight if needed, getting enough exercise, and coping more effectively with stress.

The doctor may prescribe one or more types of medications to treat CAD. These medications may prevent or delay the need for surgery:

- Drugs to lower blood cholesterol levels. These may include niacin, fibrates, and a group of drugs known as statins.
- Aspirin and other drugs that prevent blood clots from forming.
- Nitroglycerin. This drug relieves the pain of angina by opening (dilating) the coronary arteries and by reducing the heart muscle's need for oxygen.
- Beta blockers. These drugs slow down the heart rate, lowering blood pressure and reducing the heart's need for oxygen.
- ACE inhibitors. Drugs in this group dilate the coronary arteries and lower blood pressure.
- Calcium channel blockers. These drugs also dilate the coronary arteries to increase the flow of blood to heart muscle.

In some cases the patient's doctor may recommend surgery in order to prevent a fatal heart attack. The two operations that are most commonly performed are coronary artery bypass surgery and coronary angioplasty. In bypass surgery, the surgeon takes a piece of a healthy artery from another part of the patient's body and sews it in place to go around a blocked coronary artery to restore normal blood flow to the heart. In a coronary angioplasty, the surgeon inserts a catheter with a special balloon tip into the coronary artery. When the catheter is in the proper position, the balloon is expanded, which reopens the blocked artery. The surgeon will then insert a stent, which is a tube made of metal mesh, to keep the artery open. Some stents also contain a slowrelease medication.

#### Prognosis

The prognosis of CAD depends on the patient's age and the number of risk factors in their family history, personal medical history, and lifestyle. The factors that doctors take into account in evaluating an individual patient's prognosis include the number of coronary arteries that are damaged; the patient's capacity for physical exercise or activity; a history of previous heart attacks or bypass surgery; and the severity of the patient's present symptoms—particularly unstable angina or a heart attack.

## Prevention

People who are at risk of CAD or been diagnosed with it can lower their risk of a heart attack by:

- Quitting smoking or not starting in the first place.
- Getting regular medical checkups. This precaution is important because risk factors for CAD like high blood pressure, high cholesterol levels, and diabetes have no symptoms in their early stages.
- Avoiding using cocaine and drinking large quantities of alcohol.
- Keeping one's weight at a healthy level and getting regular exercise.
- Controlling blood pressure.
- Learning how to manage emotional stress.
- Eating a low-fat diet rich in fruits and vegetables.

## The Future

Coronary artery disease is expected to be a growing problem in the developed countries over the next several decades because of the aging of the general population, the increase in obesity and the rates of obesity-related diseases like diabetes, and the high rates of smoking in many countries. In terms of prevention and treatment, new drugs to treat heart attacks are currently being studied as well as the effectiveness of using bone marrow or stem cells to help repair injured heart tissue. Some researchers are also looking at some genes on chromosome 17 as a possible risk factor in CAD.

**SEE ALSO** Alcoholism; Diabetes; Heart attack; Heart failure; Hypercholesterolemia; Hypertension; Sleep apnea; Smoking

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## WORDS TO KNOW

- **Angina:** Chest pain caused by an inadequate supply of oxygenated blood to the heart muscle.
- Atherosclerosis: Stiffening or hardening of the arteries caused by the formation of plaques within the arteries.
- **Cholesterol:** A fatty substance produced naturally by the body that is found in the membranes of all body cells and is carried by the blood.
- **Ischemia:** Loss of blood supply to a tissue or organ resulting from the blockage of a blood vessel.

- **Myocardial infarction:** The medical term for a heart attack.
- **Plaque:** A deposit of cholesterol and dead white cells along the inside wall of an artery.
- **Platelet:** A small flat disk-shaped body in human blood that helps to form blood clots by sticking to other platelets and to damaged tissue at the site of an injury.
- **Stem cell:** A type of unspecialized cell that can reproduce itself and differentiate into different types of specialized cells. Stem cells act as a repair system for the body.

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# m

## Creutzfeldt-Jakob Disease

#### Definition

Creutzfeldt-Jakob disease (CJD) is a rapidly progressive and invariably fatal brain disease that is caused by a prion, an infectious protein particle. There are two major forms of CJD, a so-called classic form that
is most common in adults between the ages of forty-five and sixty-five, and variant CJD (vCJD), which primarily affects younger adults in their mid- to late twenties who live in Europe, almost entirely in England.

### Description

Classic CJD is the most common of the transmissible spongiform encephalopathies (TSEs) found in humans. These diseases are called transmissible because people (and animals) can get them through blood transfusions, tissue transplantation, or eating infected food materials. Spongiform means that the brain of a person or animal that has died from a TSE has microscopic holes in it at autopsy and a generally spongy texture when examined under a microscope. Encephalopathy is the medical term for a disease of the brain that causes changes in the function of the brain. CJD is named for the two German doctors who first described it, Hans Gerhard Creutzfeldt (1885–1964) and Alfons Maria Jakob (1884–1931).

CJD is related to several other rare prion diseases, including kuru, a disease that was epidemic in Papua New Guinea in the mid-twentieth century; and fatal familial insomnia (FFI), an inherited disease that has been identified in only twenty-eight families around the world.

Other prion diseases infect animals; these include so-called mad cow disease and scrapie, a disease found in sheep and goats. Some think that mad cow disease is a form of scrapie and may have started in the early 1980s, when the remains of dead sheep were used as a protein supplement in cattle feed. The importance of mad cow disease is its relationship to variant CJD (sometimes called new variant CJD), the other major human form of TSE. Variant CJD is thought to result from eating meat from a cow infected with mad cow disease just as mad cow disease resulted from feeding protein derived from infected sheep or cattle to cattle. This type of feeding process is no longer used.

### **Demographics**

Classic CJD and variant CJD are both very rare diseases. Classic CJD occurs in about one person in every 1 million per year around the world. There are about 200 cases reported each year in the United States.

Variant CJD (vCJD) was first reported in the United Kingdom in 1995. Of the about 200 known or suspected cases of vCJD, 164 were reported in the United Kingdom, twenty-one in France, four in Ireland,

### Also Known As

CJD, classic CJD, variant CJD, transmissible spongiform encephalopathy (TSE)

### Cause

Prion (infectious protein particle)

### Symptoms

Long incubation period, then memory loss, hallucinations, and rapid progression to death

### Duration

Death within five to fourteen months of symptom onset



Brain of a patient who died from CJD (new variant). The green areas in the center are damaged by the disease. SPL/ PHOTO RESEARCHERS, INC.

and three in the United States. Of the three cases reported in the United States, two of the patients had been born in the United Kingdom and lived there for some years; the third had grown up in Saudi Arabia. As of 2008, there have been no cases of vCJD that have originated in the United States.

CJD is thought to be equally common in both sexes and all races.

### **Causes and Symptoms**

CJD is thought to be caused by a prion, an infectious protein particle that causes similar normally folded protein molecules in the brain to refold themselves into abnormal shapes. The misfolded proteins accumulate in

### Death of a Master Choreographer

George Balanchine (1904–1983), a noted dancer and choreographer, left his native Russia for England in 1924 and emigrated to the United States in 1933. A dance company that he founded after his arrival became the New York City Ballet in 1948. His 1954 staging of *The Nutcracker* helped to make the ballet an annual Christmastime tradition in the United States.

In addition to choreographing ballets for members of his company, Balanchine continued to dance himself. In 1978, however, he began to lose his balance while practicing; then he began to lose his eyesight and hearing. By 1982 he was completely incapacitated. It was only after his death in 1983 that his doctors recognized that he had CJD, but how or where Balanchine could have been infected is unknown.

One doctor who treated Balanchine in his last years implicated treatments Balanchine received to stay young. The doctor said, "Balanchine was very much concerned about staying as youthful as possible.... He once told me that in the past he had obtained 'rejuvenation' injections in Switzerland.... Such injections ... oftentimes contain



*George Balanchine.* COURTESY OF THE LIBRARY OF CONGRESS.

extracts of animal glands.... If we were certain of how and where Mr. Balanchine got this rare and fatal disease, it would help prevent others from being inoculated with it."

the brain tissue, interfere with the functioning of the brain cells, and eventually cause the death of the brain tissue. Classic CJD is thought to have an incubation period as long as twenty to forty years. A person can develop CJD in one of three ways:

- Sporadic. Sporadic means that the disease appears at random for no apparent reason. About 85 percent of cases of CJD are sporadic.
- Genetic. A gene mutation in some families accounts for 5–10 percent of cases of CJD.

 Acquired. CJD can be acquired through blood transfusions or transplantation of tissues taken from a person with CJD. This is the rarest form of CJD, accounting for about 1 percent of known cases. It is important to know, however, that people cannot get acquired CJD through the air or from casual contact with a CJD patient. It is exposure to brain or spinal cord tissue or spinal fluid from an infected person that puts others at risk.

The chief difference between classic CJD and variant CJD is the age group affected. Most persons with classic CJD are middle-aged or older adults, whereas the average age of persons with variant CJD is twentyeight years. It is also thought that variant CJD may have a shorter incubation period than classic CJD, perhaps only eleven or twelve years in length.

The symptoms of CJD include psychiatric as well as physical symptoms:

- Anxiety and nervousness
- Difficulty walking normally; stumbling and falling
- · Loss of coordination and spontaneous muscle twitching
- Seizures
- Visual problems and eventual blindness
- Mental confusion and personality changes
- Rapidly developing delirium and dementia
- Difficulty talking normally
- Hallucinations

### Diagnosis

A definite diagnosis of CJD can be made only after the patient has died and a sample of brain tissue is examined. Prior to death, however, doctors can make a tentative diagnosis by first of all ruling out certain other diseases that can cause similar symptoms. They can give the patient a spinal tap in order to rule out some other diseases. A CT scan can be performed to rule out stroke. The most important diagnostic tests are an electroencephalogram, or EEG, which measures brain waves; and a magnetic resonance imaging test, or MRI. Patients with CJD will have a specific type of abnormal brain wave pattern that is not found in any other disorder. MRI images often reveal patterns of damage to brain tissue that are characteristic of CJD.

### Treatment

There is no known treatment that will cure CJD. Researchers have tried a variety of medications, including steroids, antibiotics, antiviral drugs used to treat AIDS, and several other experimental drugs. Treatment consists of making the patient as comfortable as possible. Morphine and similar drugs can be given to relieve pain. Some other anti-seizure drugs may be given to prevent seizures. The patient can be turned frequently to prevent bedsores and be given fluids and nutrition intravenously.

### Prognosis

CJD is a rapidly progressing disease with a fatal outcome. Most people develop dementia within six months of the onset of symptoms and become completely unable to take care of their basic physical functions shortly afterward. Most of those with classical CJD die within seven months; however, a few patients live as long as one to two years after diagnosis. Death is usually caused by infection, heart failure, or respiratory failure. Those with variant CJD may survive longer, perhaps a year or two.

### Prevention

There is no known way to prevent genetic transmission of classic CJD, as patients usually are old enough to marry and start a family long before the symptoms of the disease appear. There is also no known way to prevent the sporadic form of the disease. Some public health measures, however, have been put into place:

- People with suspected CJD or with a family history of the disease are discouraged from being organ or tissue donors.
- Health care workers are required to wear gloves and face masks when handling body fluids from a patient with CJD or when caring for the patient's wounds.
- Surgical instruments that have touched the patient are sterilized by soaking for a full hour in undiluted chlorine bleach and then heated in distilled water in an autoclave (pressure cooker) for an hour at 270–273°F (132–134°C).
- Standards for strict management of infected cows and restrictions as to what they are fed.

### WORDS TO KNOW

**Dementia:** Loss of memory and other mental functions related to thinking or problem-solving.

- Fatal familial insomnia (FFI): A very rare inherited prion disease in which the person dies of sleeplessness.
- Kuru: A fatal brain disease related to CJD that was epidemic in Papua New Guinea in the mid-

1950s. Kuru is thought to have been spread by cannibalism.

**Mad cow disease:** A prion disease that affects cattle and can be transmitted to humans by eating meat from infected cattle.

Prion: An abnormal infectious protein particle.

Scrapie: A prion disease of sheep and goats.

### The Future

Researchers continue to look for a cure for CJD. One of the major problems is the nature of prions themselves; unlike bacteria or viruses, they are very difficult to kill and do not contain any genetic material (DNA or RNA). A better understanding of the nature of prions may lead eventually to an effective treatment for CJD.

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# **u** Crohn Disease

### Definition

Crohn disease is a chronic (long-term) disease of the digestive tract marked by severe inflammation. It may involve any part of the digestive system from the mouth to the rectum, but is most likely to affect the small intestine and the area around the anus. Crohn disease is similar to ulcerative colitis, another disease of the digestive system that is sometimes categorized together with Crohn disease as inflammatory bowel disease (IBD). The major difference between ulcerative colitis and Crohn disease is that ulcerative colitis is usually restricted to the large intestine and involves the upper layers of the bowel lining, while Crohn disease can occur anywhere in the digestive tract and affects deeper layers of tissue.

Crohn disease is named for Burrill Bernard Crohn (1884–1983), a gastroenterologist (doctor who specializes in diseases of the digestive system) who treated patients at Mount Sinai Hospital in New York City.

### Description

Crohn disease is a chronic inflammatory disorder of the digestive system that is thought to be caused by an abnormal response of the body's immune system to bacteria in the digestive tract. A gene was identified in 2006 that may also be linked to the disease. Crohn disease is marked by breaks in the lining of the digestive tract, commonly in the small intestine, that cause cramps, nausea, persistent diarrhea, fever, and rectal bleeding. The symptoms vary in severity from patient to patient; some people may have only occasional episodes of diarrhea, for example, while others may have twenty to thirty bowel movements in a single day that

### Also Known As

Regional enteritis, inflammatory bowel disease, IBD

### Cause

Genetic factors combined with immune system problems

### Symptoms

Stomach cramps, pain in intestines, diarrhea, weight loss, nausea, bloody stools

#### Duration

Lifelong after diagnosis

Crohn Disease

interfere with sleep, work, school, or other activities. In addition, it is not unusual for patients with Crohn disease to have sections of normal healthy bowel alternating with sections of diseased bowel tissue.

In one type of Crohn disease, sometimes called penetrating Crohn disease, the underlying layers of intestinal tissue are damaged also, leading to perforation (puncturing) of the intestinal wall. This may cause a serious infection in the abdomen or the formation of fis-

tulas, abnormal passageways leading from one loop of the intestine to another or from the abdomen to the outside of the body. These fistulas, present in about 30 percent of patients, are most likely to form in the area around the anus, leading to the formation of abscesses.

Another subtype of Crohn disease is called stricturing disease. Stricture is the medical term for an abnormal narrowing of a hollow organ such as the bowel. The inflammation and swelling of tissue inside the bowel leads to changes in the size of the patient's stools and eventual blockage of the intestinal passages. Severe abdominal cramping is often an indication of stricturing disease, as are nausea and vomiting.

In addition to intestinal problems, Crohn disease can produce long-term complications including skin ulcers, mouth ulcers similar to canker sores, eye disorders, inflammation of the liver, and joint pains or arthritis.

### Demographics

It is estimated that there are about 500,000 persons with Crohn disease in the United States, with another 500,000 suffering from ulcerative colitis. Another statistic given by some doctors is seven cases per 100,000 in the general population in Canada and the United States. Crohn is primarily a disorder of adults, most often beginning in late adolescence or the early adult years. The most common age at onset is between fifteen and thirty years, although the disorder may begin at any age.

The rate of Crohn disease in North America has been increasing since the 1960s, although the reasons for the increase are unknown. Southern Europe, South America, Africa, and Asia have considerably lower rates of the disease—as low as 0.5–0.08 cases per 100,000 people.

View of the inside of a person's intestine. The patient with Crohn disease has a pattern of small bumps on the normally smooth wall. © PHOTOTAKE INC. / ALAMY.



### A Presidential Emergency

Many people know that President Dwight D. Eisenhower (1890–1969) had a heart attack during his first term in the White House. What is less well known, however, is that Eisenhower also suffered from the stricturing form of Crohn disease. In the early hours of June 8, 1956, the president awoke with vague pains in his lower abdomen that gradually got worse over the next hours. The surgeons were concerned about the risks of an operation because Eisenhower had his heart attack just nine months before.

In spite of the risks of surgery, the president's doctors decided that an untreated bowel obstruction was even more dangerous. They discovered when they opened Eisenhower's abdomen that he did indeed have a stricture from Crohn disease that could have killed him. The surgeons bypassed the obstruction and reconnected healthy portions of the president's intestines. Ike was back at his desk conducting official business only five days after the surgery.



*Dwight D. Eisenhower*. COURTESY OF THE NATIONAL ARCHIVES AND RECORDS ADMINISTRATION (NARA).

Around the world, however, the rates of Crohn disease are higher in cities than in rural areas and higher among people with higher incomes than among lower-income groups.

One argument for suspecting a genetic factor in Crohn disease is that it runs in some families; people with siblings who have the disease are thirty times more likely to develop it than the normal population. It is also relatively common among certain ethnic groups, particularly Jews of Eastern European origin. A two- to four-fold increase in the frequency of Crohn disease has been found among the Jewish population in the United States, Europe, and South Africa compared to other ethnic groups.

In terms of other ethnic groups in the United States, Crohn disease appears to be slightly more common in non-Jewish Caucasians than in African or Asian Americans. The disease is almost twice as likely to occur in men than in women.

### **Causes and Symptoms**

The causes of Crohn disease are not completely understood. It is known that smoking is a risk factor for developing Crohn. At one time it was thought that the disease was caused by emotional stress or by diet, particularly by eating sweet or high-fat foods. The most common theory, however, holds that the disease is caused by the patient's immune system mistaking bacteria that normally live in the intestine, and possibly certain foods, as foreign substances that must be attacked. When the immune system overreacts, white blood cells move in large numbers to the intestines, where they accumulate and eventually cause swelling and destruction of tissue.

In addition to the symptoms that have already been described, patients with Crohn disease may experience:

- · General lack of energy
- Loss of appetite
- Intestinal gas
- Nutritional deficiencies resulting from poor absorption of nutrients from food
- In children, delayed growth

### Diagnosis

No single test can be used to diagnose Crohn disease. In addition to blood tests to check for anemia and stool tests to check for blood in the stool, the four tests most commonly used to diagnose Crohn are barium studies, computed tomography (CT) scans, sigmoidoscopies, and colonoscopies. In a barium study, the patient is given barium in enema form to coat the lining of the colon and rectum. Air is then blown into the colon in order to fill it. The resultant x-ray can detect abnormalities in the lining of the intestine. CT scans are useful in detecting fistulas and abscesses.

Sigmoidoscopies and colonoscopies require special equipment inserted into the patient's body. A sigmoidoscope is a flexible lighted tube that can be inserted into the rectum and used to examine the last 2 feet (0.6 meters) of the colon. This procedure can be done in a doctor's office but does not provide a view of the entire colon. A colonoscope is a long flexible tube attached to a video camera and monitor that allows the doctor to examine the entire length of the patient's colon and rectum. The patient must take a laxative the night before to cleanse the bowel and may be given a sedative in the doctor's office to make them more comfortable. The doctor can take tissue samples from the lining of the bowel for analysis.

### Treatment

There is no medical or surgical cure for Crohn disease. Treatment consists of managing the patient's symptoms, getting the disease into remission, and preventing relapses.

Patients may be given one or more different types of medications to relieve pain and discomfort including cortisone and other drugs that reduce inflammation; drugs that block or lower the body's immune response; antidiarrheal drugs and fluid replacements; antibiotics; and nutritional supplements. Special high-calorie liquid formulas may be prescribed. Although doctors no longer think that diet causes Crohn disease, they usually advise patients who are having a flare-up to avoid bulky grains, spicy foods, alcohol, and milk products until their symptoms diminish.

Patients who are not helped by medications or who have structuring Crohn disease are usually treated by surgery. In most cases the surgeon removes the diseased part of the intestine and reconnects the healthy portions. This procedure may have to be repeated, however, if inflammation develops in the area of the intestine next to where a diseased portion was removed. In cases in which the disease is located in the large intestine (colon), the surgeon may have to remove the entire colon in a procedure called a colostomy. In this procedure, an opening called a stoma is made in the wall of the abdomen, and a portion of the remaining colon is attached to the stoma. The person's body wastes pass through the stoma and are collected in a special bag attached to the outside of the body.

Another type of surgery for patients with stricturing Crohn disease involves widening the intestine at the point of the stricture.

### Prognosis

Most people with Crohn disease have periods of remission and are able to hold jobs and generally lead normal lives. Medical treatment of Crohn disease, however, becomes less effective over time; about 80 percent of patients require surgery eventually. In addition, the disease can recur after surgery. The chance of a shortened life span or serious complications increases with the duration of the illness; patients with Crohn disease

### WORDS TO KNOW

- **Abscess:** A pus-filled sore surrounded by inflamed tissue.
- **Fistula:** An abnormal tunnel or passage that forms between one part of the intestine and another or between the intestine and the body surface.
- **Gastroenterologist:** A doctor who specializes in diagnosing and treating diseases of the digestive system.
- **Remission:** A period in the course of a disease when symptoms disappear for a time.
- **Stoma:** An opening made in the abdomen following surgery for colon cancer that allows wastes to pass from the body.
- **Stricture:** The medical term for an abnormal narrowing of a hollow organ like the bowel.

have an increased risk of colorectal cancer. The disease itself, however, is rarely fatal.

### Prevention

There is no known way to prevent Crohn disease, because its causes are not yet understood.

### The Future

It is possible that the incidence of Crohn disease will continue to rise as it has for the past fifty years. New treatments and new diagnostic techniques for the disease are currently at the clinical trials stage; as of spring 2008, the National Institutes of Health was conducting over 200 separate trials of new drugs and other types of treatment for the disorder.

SEE ALSO Canker sores; Colorectal cancer; Irritable bowel syndrome

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## **Crossed Eyes**

See Strabismus.



## **Cystic Fibrosis**

### Definition

Cystic fibrosis, or CF, is a fatal inherited disease caused by a mutation in a gene that leads to the buildup of thick, sticky mucus in the lungs and pancreas. The mucus makes breathing difficult and prevents proper nutrition by blocking the flow of digestive enzymes from the pancreas. Most patients eventually die in late adolescence or their early adult years from damage to the lungs caused by the buildup of scar tissue (fibrosis).

### Description

CF is a disease that affects many organ systems in the body. The defective gene involved in cystic fibrosis results in lower levels of chloride, a

### Also Known As

CF, fibrocystic disease of the pancreas, mucoviscidosis

### Cause

Mutation in the CFTR gene

### Symptoms

Highly salty sweat, sticky mucus blocking lungs and pancreas, death in midlife

### Duration

Lifelong



Image of a section of cystic fibrosis sufferer's lungs. The bronchial tubes are clogged with excess mucus. CNRI / PHOTO RESEARCHERS, INC.

chemical compound that is important in the production of sweat, digestive juices, and mucus of normal consistency. Chloride regulates the flow of water in and out of cells, which is needed to produce mucus that is thin enough to act as a lubricant and sweat that contains normal amounts of salt. When there is not enough chloride, the mucus that the tissues produce is thicker and stickier; it interferes with the proper functioning of the lungs, pancreas, sweat glands, and digestive tract. The thick mucus in the lungs provides a breeding ground for bacteria and fungi, which cause inflammation and gradual destruction of lung tissue.

In the digestive system, the mucus blocks the ducts that convey digestive enzymes from the pancreas, a large gland located just below the stomach. Without these enzymes, the patient's body cannot absorb nutrients from food as it passes through the digestive tract, a condition known as malabsorption. The thick mucus may also cause irreversible damage to the pancreas itself and cause inflammation of the liver as well. In men, it may block the tubes that carry sperm from the testicles, thus leading to infertility.

### **Demographics**

Cystic fibrosis is primarily a disease of people of northern European ancestry; in fact it is the most common fatal inherited disease among Caucasians in the United States, affecting one child in every 3,200. CF

### Tay-Sachs Disease (TSD)

TSD is another fatal inherited disease. It is caused by a genetic mutation in the HEXA gene on chromosome 15. It occurs in one in every 320,000 newborns in the United States. A normal HEXA gene helps to produce an enzyme that prevents a fatty substance called GM2 ganglioside from building up in the nerve cells of the brain. Without the enzyme, the child's brain cells begin to deteriorate and die. Children with TSD seem to develop normally until they are about six months old. They then begin to lose their mental functions. By age two, most children with TSD develop seizures, become blind, paralyzed, and unable to respond to their parents. They usually die around age five. As with cystic fibrosis, there is no cure for TSD.

TSD is inherited in the same pattern as cystic fibrosis; that is, both parents must have an abnormal HEXA gene for their child to be born with Tay-Sachs. About one person in every 250 in the general American population is a carrier of TSD. However, the defective gene is much more common among Jews of Eastern European descent (one in every twenty-seven) and French Canadians (Cajuns) living in Louisiana (one in every twenty-five). affects one in every 9,200 Hispanic children, one in every 15–17,000 African American children, and one in every 31,000 Asian American children. As of 2008, there were about 30,000 people with CF in the United States and about 70,000 in the world.

Males and females are equally likely to be born with CF; however, females are more severely affected and tend to die at younger ages.

### **Causes and Symptoms**

Cystic fibrosis is caused by a mutation in the CFTR gene located on chromosome 7. The gene itself was first identified in 1989. A child must inherit the defective gene from both parents in order to develop CF. If either parent has a normal CFTR gene, the child may be a carrier for the disease but will not have any symptoms of cystic fibrosis. There are over 900 different known mutations of the CFTR gene. There are an estimated 10 million carriers of the defective gene in the United States, or almost one in every thirty-one people. If two carriers marry, they have a 25 percent chance of having a child with cystic fibrosis, and a 50 percent chance of having a child who will be a carrier of the defective CFTR gene.

Some mutations of the CFTR gene cause more severe symptoms than others or may affect

different body systems differently. In addition, the symptoms of CF vary somewhat with age. In newborns, the intestines may be blocked by meconium (a blackish-green stool that most babies pass in the first few days after birth). These children may also have such symptoms as failure to grow; bulky and greasy stools; and frequent respiratory infections.

In older children and young adults, symptoms of CF may include:

- An extremely salty taste to the skin.
- Constipation or blockage of the bowels.
- Frequent episodes of heartburn or indigestion.

- Smelly, greasy, or clay-colored stools.
- Coughing or wheezing.
- Frequent chest infections with bronchitis or pneumonia.
- Clubbing (swelling of the tips) of the fingers and toes.
- Failure to gain weight at a normal rate. Girls may have delayed puberty.

Some complications of CF that occur in some patients include liver damage; polyps (tissue growths) in the nose caused by recurrent sinus infections; frequent headaches; pain in the abdomen; and diabetes resulting from damage to the pancreas.

### Diagnosis

It is possible to diagnose CF before birth around the eleventh week of pregnancy by testing a small piece of tissue from the placenta, the temporary organ that allows for exchange of nutrients and waste products between the mother and baby. After the sixteenth week of pregnancy, the fetus can be tested by amniocentesis, a procedure that involves withdrawing a small amount of fluid from the sac that surrounds the baby before birth.

Many hospitals have newborn screening programs to evaluate babies for CF. Although the screening methods are not diagnostic tests in the strict sense, they can be used to rule out CF in most babies. In addition, by identifying babies who should be tested further for CF, screening allows treatment for the disease to be started as early as possible.

The sweat test has been considered the best diagnostic test for cystic fibrosis for over forty years. It measures the amount of chloride in the person's sweat. To perform the test, the doctor applies a chemical that causes sweating on a small area of the person's arm or leg. An electrode is attached to the treated area and a mild electrical current is used to stimulate sweating. The patient may feel a tingling or warm sensation. After about five minutes, the sweat from the treated area is collected on a piece of filter paper and sent to a laboratory for analysis. One benefit of the sweat test is that the values are not changed as a person grows older or affected by temporary illness.

Another more recent test that can be used to diagnose CF is genetic testing of a sample of the patient's blood for mutations in the CFTR gene.

### Treatment

There is no cure for CF. Treatment is aimed at thinning the mucus secretions, keeping the patient's airways clear, preventing infections, and maintaining adequate nutrition.

Medications used to treat CF include:

- Mucolytics. These are drugs that thin the mucus in the lungs so that it is easier for the patient to cough it up. Pulmozyme is the most commonly used drug of this type.
- Antibiotics. These are given to treat bacterial infections in the lungs and throat. Some antibiotics can be given in aerosolized form and delivered directly into the airway.
- Bronchodilators. Bronchodilators are drugs that help to keep the bronchi (tubes leading into the lungs) open to ease the patient's breathing. They can be taken through inhalers or nebulizers.
- Oral enzymes and vitamin supplements. Patients with CF need to take pancreatic enzymes in the form of pills or capsules with their meals in order to absorb nutrients from their food. Extra vitamins are also needed to prevent malnutrition.
- Pain relievers. Some children with CF are helped by taking ibuprofen (Advil or Motrin) for headaches. This particular non-aspirin pain reliever also may help slow down loss of lung function.

Adults as well as children with CF usually need to follow a special high-calorie high-fat diet to maintain good health. A professional dietitian can work with the patient to include foods that the patient enjoys eating as well as meeting the calorie and fat requirements.

Patients with CF also need to clear the mucus from their lungs at least twice a day. This clearing can be done by having the patient lie with his or her head over the edge of a bed and thumping or clapping on the back and chest for twenty to thirty minutes to shake the mucus loose so that the patient can cough it up. As an alternative to having a physical therapist or trained family member thumping the patient's chest, the patient can wear a device called a ThAIRapy Vest, which contains a compressor that vibrates the patient's chest wall at set times to loosen the mucus. The vest costs about \$16,000.

CF patients with advanced lung disease may benefit from lung transplantation.

### Prognosis

Cystic fibrosis is a fatal disease. The average life expectancy of patients is 36.8 years, with males on average living longer than females. This is a dramatic increase over the 1950s, when few patients with CF lived long enough to start elementary school. About 80 percent of patients live to be adults; a few patients live into their forties. Most patients die of end-stage lung disease.

Most males with CF (97 percent) cannot father children. Women with CF are more likely to be fertile; however, they must watch their diet very carefully during pregnancy and may find that pregnancy worsens the symptoms of CF.

### Prevention

There is no way to prevent cystic fibrosis other than genetic testing of potential parents. Genetic screening of family members of a cystic fibrosis patient may detect the CF gene in between 60 and 90 percent of carriers, depending on which test is used. There are still a few rare mutations of the CFTR gene that cannot be identified by present tests.

### The Future

Cystic fibrosis was the first disease that researchers attempted to treat by using gene therapy, a technique for replacing defective genes in the body's tissues with normal genes. In 1993, scientists used a common cold virus in an attempt to deliver a healthy CFTR gene to the cells in the patient's airways. Although this first attempt was not successful, other methods of gene delivery are now being tested, including nose drops, fat capsules, and solutions delivered by nebulizers.

Another avenue of research involves mapping the genome of the bacterium that causes most of the lung infections in CF patients. By cracking the bacterium's genetic code, researchers are hoping to develop better drugs for treating the infections that it causes.

SEE ALSO Bronchitis; Pneumonia

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### WORDS TO KNOW

- **Bronchus (plural, bronchi):** One of the two major divisions of the airway that lead into the right and left lungs.
- **Clubbing:** Swelling of the tips of the fingers. Clubbing is one of the symptoms of CF.
- **Fibrosis:** The medical term for the formation of scar tissue.
- Gene therapy: The insertion of normal genes into a person's cells or tissues in order to treat a

disease by replacing a harmful mutation or nonfunctioning gene.

- **Malabsorption:** Inability to absorb the nutrients in food through the digestive tract.
- **Meconium:** A dark greenish type of stool passed by a newborn during the first few days of life. Inability to pass the meconium is an early symptom of CF.
- **Nebulizer:** A device that delivers medication in a fine spray or mist.

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# D



### Also Known As

Depressive disorder, dysthymia

### Cause

Genetic, biochemical, and environmental factors

### Symptoms

Depressed mood; sleep disturbances, crying spells, weight loss or gain, thoughts of suicide

### Duration

Months to years; may come and go

### Deafness See Hearing loss.



## Depression

### Definition

Depression, sometimes called major depression or depressive disorder, is a mood disorder that has been called the "common cold of mental illness" because it is so common in the general population. Unlike the common cold, however, depression can have serious long-term effects on a person's quality of life.

Some types of depression are recognized as separate disorders:

- Dysthymia. Also called dysthymic disorder, dysthymia is a condition in which the person has milder symptoms of depression that are less disabling than those of major depression over a period of two years or longer.
- Psychotic depression. Patients with this type of depression have hallucinations, delusions, and other signs that they have lost contact with reality.
- Postpartum depression. This is a type of depression that affects some women following the birth of a baby.
- Seasonal affective disorder (SAD). SAD is a form of depression related to changes in the seasons. Most people with SAD feel

depressed in the winter and better in summer, but there is also a reverse form of SAD in which the person is depressed in summer and feels better in winter.

### Description

Depression is a mood disorder that does not affect everyone in the same way. There is some evidence, for example, that women and men experience depression differently; women are more likely to feel sad, worthless, or guilty, while men are more likely to feel tired, irritable, and uninterested in activities that they used to enjoy. Men appear more likely than women to get angry and act abusively toward others, or to drive recklessly. Depression in the elderly may take the form of memory problems or slowed-down movement rather than crying or feeling sad. Depression in children may have such symptoms as refusing to go to school, getting in trouble, or sulking and being generally moody and hard to get along with.

Depression also has different patterns of recurrence in different people. Some persons have one episode of depression, recover, and are never troubled by recurrences. Others have a series of episodes at irregular intervals. And as the definition of dysthymia indicates, some people have a low-grade depression that may persist for years without turning into an episode of major depression.

Depression often coexists with other mental and physical disorders, which often makes it difficult to diagnose. Depression can occur before the person gets sick; can occur as a result of the sickness; or exist alongside other illness. Mental disorders that often coexist with depression include alcoholism and substance abuse disorders, eating disorders, anxiety disorders, posttraumatic stress disorder, and obsessive-compulsive disorder. Physical disorders that commonly affect depressed patients include heart disease, stroke, cancer, AIDS, diabetes, and Parkinson disease. Depression can make the symptoms of these diseases worse and harder to treat.

### Demographics

Depression is one of the more common mood disorders in the United States. According to the National Institutes of Health, one in every five women and one in every eight men will have an episode of major depression at some point in their lifetime. Depression is a major cause of time lost from work, lost opportunities for education, and vulnerability to substance abuse. It is a factor in 55 percent of all suicide attempts, or about 110,000 attempts annually. Depression adds to the cost of treating such physical illnesses as diabetes, heart disease, high blood pressure, and cancer.

People can suffer from depression at any age; however, adults between the ages of thirty and forty are most likely to be diagnosed with major depression. There is a second but smaller peak in adults between fifty and sixty.

- Major depression is diagnosed twice as often in women as in men; however, in children, boys are diagnosed with depression as often as girls.
- Depression appears to be less common among African Americans in the United States than among members of other racial groups. Risk factors for depression in adult life include:
- Death of a parent during one's childhood
- A family history of depression
- A history of suicide in the family
- Long-term use of certain medications, particularly birth control pills, drugs given to treat high blood pressure, and sleeping pills
- Long-term alcohol or drug abuse
- Poverty and unemployment
- Recent bereavement or traumatic incident

### **Causes and Symptoms**

The causes of depression have been debated for decades, with researchers disagreeing as to whether biology, psychology, or a combination of the two offers the best explanation. Most researchers now think that depression is the end result of biological vulnerability to a mood disorder combined with personal history and certain personality traits.

- Genetic factors. No specific genes have been identified, although the disorder is known to run in families.
- Biochemical. Researchers have found that the brains of people with depression have abnormal levels of certain brain chemicals called neurotransmitters. Neurotransmitters relay impulses from one nerve cell to the next.
- Life history. Difficult circumstances early in life, too many traumatic experiences too close together, or high stress levels over a long period of time can all make people more likely to become depressed.
- Personality factors. People who are pessimistic, easily discouraged, or inclined to worry a lot are vulnerable to depression.

An occasional blue mood or temporary feeling of discouragement is not a depression. For a diagnosis of depression a person must have five symptoms from the following list for at least two weeks. The symptoms must be severe enough to interfere with the person's daily activities and relationships:

- Depressed mood
- Loss of interest or pleasure in activities that the person used to enjoy
- · Weight gain or loss
- Difficulty sleeping or sleeping much more than usual
- Slowed movement or extreme restlessness
- Lack of energy; difficulty getting things done
- Feeling worthless or hopeless
- Problems with concentrating or decision making
- Thoughts of death or suicide

### Diagnosis

The diagnosis of depression is complicated and often missed, particularly in the elderly. Diagnosis begins with a complete physical examination, partly to see whether the patient has medical problems that may increase his or her risk of depression, and partly to rule out physical reasons for changes in mood. These include thyroid disorders, infectious diseases like syphilis or Lyme disease, and prescription medications that are known to affect mood. The doctor may order blood or urine tests as part of the physical examination.

Another important part of the diagnosis is taking the patient's personal and family history. This part of the patient interview often includes giving the patient the Beck Depression Inventory or another questionnaire that can be completed in the doctor's office in a few minutes. There are special questionnaires of this sort for children and adolescents.

The doctor will also listen to the way the patient talks as well as the content of what they are saying, because depressed people often talk slowly and may sound sad. The patient's facial expressions and the way they are dressed may also provide clues; a patient who is usually neat and tidy may come to the office looking poorly groomed.

Primary care doctors will usually refer their patients to psychiatrists (mental health specialists) in order to distinguish major depression from other mental illnesses, and to prescribe treatments for the depression.

### Treatment

Treatment for depression may consist of antidepressant medications, psychotherapy, electroconvulsive therapy (ECT), or a combination of these approaches.

- Antidepressant medications: These are drugs that work by affecting the levels of neurotransmitters in the brain. There are several different families of antidepressant medications, and the doctor may have to try several different drugs before finding the one that works best for the patient. The choice of antidepressant also depends on whether the patient is taking prescription drugs for other health conditions. It takes anywhere from two to eight weeks for the patient to know whether the antidepressant is working for them. Between a half and two-thirds of people with depression are helped by medications.
- Psychotherapy. There are several different approaches that are used to treat depressed people. The most common ones are interpersonal therapy and cognitive therapy. In interpersonal therapy, the person learns about the causes of depression and the social triggers in his or her life that set off depressive thoughts, together with strategies for coping with their social situation. Cognitive therapy works by teaching the patient to change his or her ways of thinking. Many people have underlying negative assumptions that affect the way they see their life, and these expectations can be challenged and changed.
- Electroconvulsive therapy (ECT). Sometimes called shock therapy, ECT is a treatment in which seizures are induced in an anesthetized patient to relieve the depression. It is thought to work by changing the levels of neurotransmitters in the brain. ECT is generally used only for depressed patients who have not been helped by medications or psychotherapy.

Alternative and complementary treatments that are sometimes used for depression include various herbal remedies, such as St. John's wort. Those interested in herbal preparations should discuss these with their doctor, however, as these preparations can interact with standard prescription drugs and have side effects just like standard drugs. Other complementary therapies include acupuncture, massage therapy, music therapy, meditation, and stress reduction techniques. These are safe, and are helpful to some patients with depression. Studies also indicate that regular exercise can be helpful in controlling symptoms of depression and anxiety.

### Prognosis

The prognosis of depression varies considerably. People who are not treated for depression often feel better within six to twenty-four months; however, episodes of depression can be shortened considerably with treatment. About two-thirds of patients treated for depression feel well enough to return to their normal activities within a few weeks. About a quarter of patients will continue to have symptoms of depression for months to years after the first episode. About 50 percent of patients treated for depression will have a second episode at some point in time; these recurrences usually respond well to treatment, however.

People who are depressed are at increased risk of suicide. About 3.4 percent of patients diagnosed with major depression eventually succeed in committing suicide.

### Prevention

People cannot change some risk factors for depression, such as their family history or their sex, but they can lower their risk by taking good care of their physical health, keeping up a strong family and friendship network, learning to cope with normal life stressors, and talking to their doctor if they are concerned about their moods.

### The Future

Depression is one of the oldest known mental disorders, having been described by physicians in ancient Egypt and China, and is likely to continue to be a common problem around the world. Present research includes trials of newer antidepressants, comparisons of standard treatments with various alternative therapies, and studies of the ways in which culture or ethnic background influences depression. Another important area of research is looking for ways to predict how patients will respond to specific antidepressant medications, so that the trial-and-error approach to finding the best drug for each patient could be eliminated.

**SEE ALSO** Bipolar disorder; Child abuse; Obsessive-compulsive disorder; Postpartum depression; Posttraumatic stress disorder; Seasonal affective disorder

### WORDS TO KNOW

**Delusion:** In medicine, a false belief that a person holds to despite evidence or proof that it is false.

**Dysthymia:** A mood disorder characterized by a long-term low-key depression.

- **Neurotransmitters:** Chemicals produced by the brain that relay nerve impulses from one nerve cell to another.
- **Postpartum depression:** A type of depression that some women experience after the birth of a baby.

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# **m** Dermatitis

### Definition

Dermatitis is the general medical term for inflammation of the skin. It includes a broad range of rashes, ranging from atopic dermatitis (eczema) to sunburn and various types of skin infections.

### Description

There are five basic types of dermatitis:

- Atopic dermatitis (AD). Characterized by itching, scaling, and swelling rash on the skin that may form oozing open sores or yellow and red pimples, the rash is most likely to affect the skin of the hands and feet, the arms, the area behind the knees, and the ankles, wrists, face, neck, and upper chest.
- Contact dermatitis. Rashes caused by touching a foreign substance. There are two major forms of contact dermatitis: allergic, caused by contact with a plant or animal substance that a person has become sensitized to; and irritant, caused by touching something that damages the skin directly. Poison ivy is an example of an allergic contact dermatitis; rashes caused by detergents or other household chemicals are examples of irritant contact dermatitis.
- Cercarial dermatitis. Commonly known as swimmer's itch, it is caused by certain parasites that live in ducks, geese, and freshwater snails. (A parasite is an organism that lives off another, larger, organism.) When a person swims in water containing these parasites, they can burrow into the skin and cause a short-lived itchy

### Also Known As

Skin rash, eczema, sunburn

### Cause

Infections, parasites, sunlight, allergens, clothing, weather

### Symptoms

Reddened, itching, cracked, or bumpy skin

### Duration

Hours to weeks; some forms may be lifelong

rash with small reddish pimples or blisters. It mostly affects the parts of the body that are not covered by a swimsuit, and does not usually affect the face, palms of the hands, or soles of the feet.

- Radiation dermatitis. The most common form of radiation dermatitis is photodermatitis or sunburn. It is characterized by redness, pain, and swelling of the affected skin following exposure to sunlight or a tanning lamp. Severe sunburn includes the formation of blisters and peeling of the affected skin, which usually begins to peel about three days after exposure and may continue for another week or so. A rarer form of radiation dermatitis, found mostly among hospital staff, is caused by repeated exposure to x-ray and other imaging equipment in hospitals.
- Seborrheic dermatitis. Found on the scalp and other areas of the face and chest that contain oil-secreting glands, it is characterized by a reddish, greasy-looking rash



on the skin and oily flakes of skin. In infants, it is called cradle cap. In adults, the most common form of seborrheic dermatitis is dandruff. *Treatment of dermatitis with lotion.* SHUTTERSTOCK.

### **Demographics**

The demographics of dermatitis depend on the type:

- Atopic dermatitis. AD is more common among children than adults; 95 percent of cases occur in children five years or younger. About 15 million people in the United States have some form of eczema. The rate in adults is about 0.9 percent. AD is slightly more common in women than in men.
- Contact dermatitis. Contact dermatitis is common in the general population, affecting fourteen people per 1,000; one health survey estimated that about 9 million visits to doctors every year are for contact dermatitis. Contact dermatitis accounts for about 9 percent

### When to See the Doctor

Most cases of sore or irritated skin are not medical emergencies and do not require specialized treatment. People with the following symptoms, however, should see a dermatologist quickly because they may be early signs of skin cancer:

- A growth on the skin that was not there before the rash or sunburn.
- A sore that bleeds, crusts over, keeps reopening, and does not heal within two weeks.
- A change in the size, color, or texture of a mole.
- A dark flat spot on the skin that is gradually enlarging.

of all visits to dermatologists in the United States. Women appear to be more likely to report contact dermatitis than men.

- Cercarial dermatitis. Cercarial dermatitis can affect anyone who swims in contaminated water; however, children are more likely than adults to develop swimmer's itch because they are more likely to wade or swim in shallow water and less likely to towel dry or rinse off after swimming.
- Radiation dermatitis. Sunburn is very common in the general population in North America. According to Skin Cancer Foundation survey, 42 percent of people reported getting sunburned at least once in the preceding year. Radiation dermatitis caused by exposure to imaging equipment is now quite rare among hospital workers because the long-term impact of radiation on skin is now better understood.
- Seborrheic dermatitis. Dandruff, the mildest form of this dermatitis, is estimated to affect about 15–20 percent of adults in the United States. It is most common in people from the late teens to the late forties, and affects men somewhat more often than women. It appears to be equally common in all races and ethnic groups.

### **Causes and Symptoms**

The causes of dermatitis vary:

- Atopic dermatitis. Most doctors think that AD results from an immune overreaction inside the body that leads to inflammation and cracked, itchy skin. The breaks in the skin then let in more allergens, irritants, and microbes that made the skin itch and burn even more. A more recent theory holds that some cases of AD are caused by a defective gene for filaggrin, a protein in the skin that normally holds in moisture.
- Contact dermatitis. Contact dermatitis can be caused by a wide range of allergens and irritants. Allergens may include rubber; the nickel and other metals used in jewelry; perfume and perfumed

soaps and creams; hair dye and cosmetics; poison ivy and poison sumac; and neomycin, a common ingredient in topical antibiotic creams. Common irritants include laundry soap, skin soaps, dishwashing detergents, silver polish, shower cleaners, household ammonia, and similar products.

- Cercarial dermatitis. Cercarial dermatitis is caused by parasites that spend part of their life cycle in the water in ponds, small streams, and along shorelines. Swimmer's itch usually appears within four to forty-eight hours of exposure to water containing the parasites.
- Radiation dermatitis. Sunburn is caused by ultraviolet (UV) radiation from the sun or a tanning lamp. In rare cases it is caused by occupational exposure to x-ray and other medical imaging equipment.
- Seborrheic dermatitis. The causes of seborrheic dermatitis are not completely understood. It is thought to result from an overreaction of the person's immune system to a fungus that lives on the scalp. Emotional stress or changes in the seasons may cause flare-ups.

### Diagnosis

A primary care doctor will usually base the diagnosis by looking at the patient's skin, together with taking a personal and family history of allergies, hay fever, and skin disorders. The patient may be referred to a dermatologist (a doctor who specializes in disorders of the skin) to rule out the possibility that the rash or skin irritation is caused by other diseases. The dermatologist may take a skin biopsy in order to exclude other causes.

Swimmer's itch can be a challenge to diagnose because there are no tests specific for the parasites that cause it, and the rash looks a lot like poison ivy or chickenpox. In many cases the only clue is that the patient went swimming within the past few days.

For contact dermatitis, the doctor may perform a patch test in order to identify the specific substance(s) causing the rash. Small quantities of suspected allergens or irritants in individual containers or patches are applied to the skin of the patient's back. The patches are covered with special nonallergenic adhesive tape for forty-eight hours and then removed; the patient's skin is examined for blisters, swelling, or other reactions to the substances in the patches.

### Treatment

Treatment for dermatitis depends on the cause of the skin rash:

- Atopic dermatitis. Treatment focuses on reducing the itching (and therefore scratching) of the rash; lowering inflammation; and preventing flare-ups. Medications include moisturizers, steroid medications, antihistamines, and drugs that affect the functioning of the immune system.
- Contact dermatitis. Mild contact dermatitis is treated with creams containing cortisone that are applied directly to the rash or with antihistamines taken by mouth. Wet compresses may also help to relieve the itching. Severe cases of contact dermatitis may require oral or injected steroid medications or antibiotics.
- Cercarial dermatitis. Swimmer's itch does not usually require a visit to the doctor. It can be treated at home with over-the-counter antihistamines, calamine lotion, or cortisone cream to relieve the itching. Bathing in water containing baking soda or an oatmeal treatment like Aveeno may also help. If the itching is severe, the doctor may order a prescription-strength cream or lotion.
- Radiation dermatitis. Most mild cases of sunburn eventually heal without special attention from a doctor. Home care may include bathing in cool (not cold) water with baking soda added to the bath water; applying aloe vera gel or other non-greasy moisturizing lotion; and taking a nonaspirin pain reliever.
- Seborrheic dermatitis. Seborrheic dermatitis of the face or chest may be treated with antibiotic, antifungal, or steroid creams applied directly to affected areas. For dandruff, the doctor will usually recommend special shampoos containing salicylic acid, tar, selenium, sulfur, or zinc, and advise the patient to wash the hair more frequently and leave the shampoo on the scalp for about five minutes.

### Prognosis

The prognosis depends on the type of dermatitis:

• Atopic dermatitis. About half of children diagnosed with AD will improve by age fifteen; the other half will have lifelong symptoms. It is unusual for a person to develop eczema for the first time after age thirty unless they are working in a harsh climate or a wet environment.

- Contact dermatitis. Even without medical treatment most cases improve within a few days to three to four weeks after exposure to the allergen or irritant has stopped.
- Cercarial dermatitis. Most cases clear up by themselves in a few days to a week without medical treatment, although home treatment with calamine or antihistamines will relieve the itching more rapidly.
- Radiation dermatitis. Most mild cases of sunburn heal without problems in the short term. Blisters that become infected usually heal completely once the infection is treated. The long-term prognosis is of greater concern, as a history of repeated sunburn increases a person's risk of melanoma (the most serious form of skin cancer).
- Seborrheic dermatitis. There is no long-term cure for this type of dermatitis; however, faithful use of the shampoos and skin treatments prescribed by the doctor usually relieves symptoms and lowers the risk of flare-ups.

### Prevention

Prevention of most forms of dermatitis consists of first identifying and then avoiding the allergen or other cause of the skin rash:

- Atopic dermatitis. Prevention of AD consists of identifying the specific substances that trigger skin reactions; avoiding contact with them whenever possible; and applying products that relieve the itching, dryness, and inflammation of the skin.
- Contact dermatitis. In addition to advising the patient to avoid the allergen or irritating substance, in some cases the doctor can recommend substitutes for the specific products or substances causing the rash.
- Cercarial dermatitis. To prevent swimmer's itch, people should choose swimming areas carefully; avoid marshy or shoreline areas where snails often live; rinse off thoroughly after swimming; and use chlorine to keep home pools free of parasites.
- Radiation dermatitis. Staying out of the sun between 10 a.m. and 4 p.m.; wearing a hat and clothing that covers as much of the body as possible; and using sunscreen when outdoors.
- Seborrheic dermatitis. Avoiding the use of hair spray or greasy hair gels or creams is usually necessary. Stress management techniques are helpful to some patients in preventing flare-ups caused by emotional stress.

### WORDS TO KNOW

Allergen: Any substance that can provoke an allergic reaction in susceptible individuals.

**Contact dermatitis:** Inflammation of the skin caused by direct contact with an allergen or

irritating substance, such as poison ivy, certain dyes, or certain metals.

**Dermatologist:** A doctor who specializes in diagnosing and treating diseases of the skin.

### The Future

Dermatitis is likely to continue to be a commonplace health problem, if only because it has so many forms and causes.

SEE ALSO Eczema; Hives; Sunburn

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# m

## **Developmental Disability**

### Definition

Developmental disability is a term applied to a group of chronic disorders caused by physical and/or mental impairments that occur before age twenty-two and typically last for a lifetime. These disorders affect a person's ability to learn, speak effectively, take care of themselves, hold a job, and live independently.

Developmental disabilities are defined as mild, moderate, severe, or profound on the basis of how much social support the person needs. A person with mild developmental disabilities, for example, may be able to benefit from special education programs, finish high school or a vocational training program, work in certain types of jobs, and live with their family or in the community. Those with profound disabilities, on the other hand, may require twenty-four-hour care in a group home or institution.

### Description

Developmental disabilities can be described as falling into one of several types:

- Disabilities involving the brain, spinal cord, and nervous system. These developmental disabilities include mental retardation (Down syndrome, fragile X syndrome) and autism spectrum disorders.
- Disabilities related to hearing or vision loss.
- Disabilities related to metabolic disorders. Metabolism refers to the sum total of the chemical changes in the body that are needed to provide energy and repair or grow tissues. Metabolic disorders

Also Known As Mental retardation, children with special needs

### Cause

Diseases, trauma, birth defects, or genetic disorders

### Symptoms

Mental retardation, speech and behavioral problems, difficulties with self-care

**Duration** Lifelong

# Developmental Milestones in Children

Here are some of the physical characteristics or abilities that doctors look for in evaluating a child's development during the preschool years:

- Three months: Baby smiles when smiled at, grasps rattle, turns head toward lights or voices.
- Six months: Baby can sit in a high chair, imitate parent's hand movements, laugh, and make sing-song noises.
- Twelve months: Baby can eat finger food, crawl, walk holding parent's hand, respond to music, say first word.
- Eighteen months: Toddler can walk without help, identify an object in a picture book, say eight to ten different words, ask for something by pointing.
- Two years: Child can open boxes, drink from a straw, have a vocabulary of several hundred words, hum or try to sing, play with other children.
- Four years: Child can eat or drink without spilling; draw simple objects; undress without help; use the toilet alone; tell the difference between the real world and imaginary worlds; speak in complete sentences with good grammar.
- Five years: Child can count to ten; has an understanding of time; can hop, climb, skip, and turn somersaults.

that affect children's development include thyroid diseases and phenylketonuria.

• Disabilities related to degenerative disorders. Degenerative disorders are those in which a child appears normal at birth and reaches some developmental milestones but then starts to lose the abilities they have attained.

Developmental disabilities have been recognized for centuries even though people did not always understand their causes. Before the eighteenth century, people suffering from mental retardation and other disabilities were often cared for by their families or in religious institutions like monasteries. After the Industrial Revolution in the nineteenth century, people with developmental disabilities were more likely to be taken from their families in childhood and placed in large asylums-some containing as many as several thousand people. Some of these institutions provided an elementary education as well as food, shelter, and clothing, but most met only the basic needs of the children and adults in their care. It was not until the 1960s that doctors and teachers began to question the effectiveness of placing people with disabilities in large institutions.

Between the 1960s and the 1990s, the United States and other developed countries moved toward individualized education and social support for people with developmental disabilities. Instead of separating these persons from the wider community, doctors and teachers now focus on each individual's gifts and talents as well as their needs, and try to place them in classroom and work settings where they can interact with others. In 2000 the U.S. Congress passed the Developmental Disabilities Assistance and Bill of Rights Act, which funds four grant programs to assist people with developmental disabilities and their families.

There are still several challenges to teaching and treating or supporting people with developmental disabilities. One is that these children and adults are vulnerable to abuse by others. They also have a higher than normal rate of mental health issues, not only because they are often hurt or abused but also because they frequently become dependent on drugs or alcohol. Third, people with developmental disabilities sometimes behave in ways that are upsetting or dangerous to others, such as hitting, screaming, kicking, being sexually inappropriate, stealing or throwing objects, or injuring themselves. Treatment of people with developmental disabilities often has to include medications or psychotherapy to help them control problem behaviors.

### Demographics

According to the U.S. Administration on Developmental Disabilities (ADD), there were about 4.5 million children and adults in the United States with developmental disorders as of 2008. Some statistics for specific disorders are as follows:

- Autism: 560,000 persons between the ages of one and twenty-one years.
- Cerebral palsy: 800,000 children and adults.
- Hearing loss: 72,000 children between the ages of six and twenty-one.
- Mental retardation: 2 million children and adults between the ages of six and sixty-four.

### **Causes and Symptoms**

Developmental disabilities can be caused by a number of different factors:

- Injury to the baby's brain before birth or during the birth process.
- Mother's lifestyle. Heavy drinking, drug use, or smoking can lead to fetal alcohol syndrome or other forms of brain damage.
- Rubella or other infectious diseases.
- Extreme prematurity.
- Child abuse.
- Genetic disorders. Down syndrome, phenylketonuria, fragile X syndrome, and some degenerative disorders are all caused by genetic mutations or an extra copy of a chromosome.
- Poor diet and health care.

Developmental disabilities are defined by the U.S. government as disabilities that affect the individual's functioning in three or more of the following areas:

- The ability to live on one's own
- The ability to support oneself financially
- The ability to learn in school
- The ability to get around on one's own, by walking, driving, or using public transportation
- The ability to use and understand spoken language
- The ability to take care of one's personal needs (dressing, eating, washing, using the toilet, doing laundry)
- Self-direction (the ability to make decisions, evaluate situations, and generally be in charge of one's life)

## Diagnosis

The diagnosis of developmental disabilities is made at different points before or after the child's birth, depending on the specific disorder. Some genetic disorders, like Down syndrome, can be diagnosed before the baby is born. Metabolic disorders like phenylketonuria can be detected by a blood test used to screen babies shortly after birth.

In other cases, developmental disabilities may be identified by the baby's doctor during periodic checkups. The doctor can observe how the child interacts with its caregiver; can test his or her vision and hearing; and see whether the child has attained specific developmental milestones. Parents who have noticed that their child does not seem to be walking, talking, or responding to them at the age at which these abilities usually appear will often consult a specialist in child development for an evaluation of the child. Some learning disabilities may not become evident until after the child starts school and may be first noticed by a teacher.

# Treatment

There is no cure for developmental disabilities. Treatment of developmental disabilities is tailored to the individual child and his or her specific type of disability. It may include one or more of the following types of treatment or care:

- Medications or surgery for specific physical or emotional problems.
- Special education programs.
- Vocational training for future employment.
- Speech and language therapy.
- Physical therapy.
- Psychotherapy and counseling for behavioral problems.
- Support services for people who are profoundly disabled. These may include nursing care as well as assistance with housekeeping, personal cleanliness, shopping, and handling money.

Although many people with developmental disabilities cannot be completely self-sufficient in all aspects of daily life, treatment is aimed at helping them achieve as much independence as possible.

# Prognosis

The prognosis for children with developmental disabilities depends on the severity of the child's specific disability; whether it is progressive (gets worse over time); and the extent to which it affects his or her ability to get along with other people, finish school or train for a job, and live independently. In general, developmental disabilities shorten a person's life expectancy by about twenty years, although improvements in technology as well as medicine are helping people with disabilities have a better quality of life as well as live longer.

# Prevention

Not all developmental disabilities can be prevented. Some measures that people can take, however, to lower their risk of having a child with developmental disabilities include:

- Genetic screening for both parents, particularly if either father or mother comes from a family with a history of genetic disorders
- Proper nutrition and health care for the mother during pregnancy
- Avoidance of drinking, smoking, and drug use during pregnancy
- Vaccination against rubella before pregnancy if the mother has not been exposed to it

# WORDS TO KNOW

Chronic: Long-term.

- **Degenerative disorder:** A type of disorder in which a child gradually loses certain abilities that he or she had acquired at an earlier age.
- **Metabolism:** The sum total of the chemical changes in living cells that are necessary to maintain life.
- **Milestone:** A physical development or accomplishment that most children reach within a specific age range.
- **Progressive:** Referring to a disease or disorder that gets worse over time.

#### The Future

Developmental disabilities are likely to become a serious concern for caregivers and health care professionals in the years ahead. One reason is that some developmental disabilities, such as autism and hearing loss, are becoming more rather than less common. Another reason is that people with developmental disabilities are living longer, which means that they will require treatment and support services for many more years than was the case in the 1960s. The needs of adults over fifty with developmental disabilities are presently being studied by policy makers as well as medical researchers.

Another rapidly expanding area of research is finding better screening instruments to identify children with developmental disabilities as early as possible. The available evidence indicates that the earlier these children are diagnosed and treated, the more likely they are to develop their full potential as they grow older.

**SEE ALSO** Asperger syndrome; Autism; Cerebral palsy; Child abuse; Down syndrome; Fetal alcohol syndrome; Fragile X syndrome; Hearing loss; Phenylketonuria; Prematurity; Rubella; Seizure disorder; Shaken baby syndrome; Spina bifida

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# U

# Diabetes

#### Definition

Diabetes mellitus, or DM, is a lifelong disorder in which the patient's body cannot regulate the levels of glucose (sugar) in the blood. The problem may be caused by too little insulin—a hormone produced by the pancreas that helps the body use the glucose properly—or the body's resistance to using the insulin that is secreted by the pancreas.

There are three major types of diabetes:

• Type 1. Type 1 diabetes, which is sometimes called insulindependent diabetes because the patient must take a daily dose of insulin in order to live, is a disease in which the person's immune system attacks the cells in the pancreas that produce

#### Also Known As

Diabetes mellitus, DM

#### Cause

Inability of the body to regulate the amount of glucose (sugar) in the blood

#### Symptoms (type 1)

Thirst, increased urination, weight loss, nausea and vomiting

#### Symptoms (type 2)

Thirst, increased appetite and urination, fatigue, blurred vision, slowhealing infections

Duration Lifelong



insulin. This type of diabetes usually develops over a short period of time, most often in children and young adults. It accounts for 5–10 percent of cases of diabetes diagnosed in the United States.

- Type 2 diabetes. Type 2, sometimes called adult-onset diabetes, is the most common type, accounting for 90–95 percent of cases in North America. It usually develops relatively slowly and is associated with being overweight and a family history of diabetes. Most people with type 2 diabetes are resistant to insulin rather than having a pancreas that is not producing enough of the hormone.
- Gestational diabetes. Some women develop a temporary form of diabetes toward the end of their pregnancy that goes away after the baby is born. Women with gestational diabetes have an increased risk of developing type 2 diabetes within five to ten years.

Doctors also consider a condition known as pre-diabetes or impaired glucose tolerance to be a health concern. People with this condition have

a higher-than-normal blood glucose level but not high enough to meet the criteria for a diagnosis of diabetes. Many people with pre-diabetes will develop type 2 diabetes within ten years.

# Description

People with type 1 diabetes are likely to feel sick fairly suddenly. The most common symptoms are increased thirst, weight loss in spite of increased appetite, nausea and vomiting, and blurred vision. If the person is not diagnosed and treated with insulin quickly, they can fall into a coma and die.

People with type 2 diabetes may not have any noticeable symptoms for some years. When they do develop symptoms, they may feel thirsty and hungry, urinate more frequently, tire easily, have vision problems, or notice that wounds and sores heal slowly. Women with gestational diabetes may not have any symptoms at all.

Diabetic taking a blood glucose test. SHUTTERSTOCK.

Diabetes is a serious disease because it often leads to complications that seem far removed from digestive issues. People with diabetes are at increased risk of blood vessel injury, stroke, blindness, heart disease, kidney failure, gangrene leading to limb amputations, and nerve damage. Women with gestational diabetes are more likely to have babies with birth defects.

## Demographics

About 21 million people in the United States, or 7 percent of the population, have diabetes. Unfortunately, 6.5 million of these people have not been diagnosed with the disease. Between 54 and 57 million Americans were thought to have pre-diabetes as of 2007. Diabetes is the sixth leading cause of death in the United States; about 65 percent of deaths among patients with diabetes are the result of heart attacks and stroke, but diabetes is the underlying cause.

Diabetes is a major cause of disability and death in the United States. It costs the country about \$135 billion each year, including \$42 billion in indirect costs (disability payments, lost work time, and premature death) and \$93 billion in direct medical costs (hospital admissions, medical care, and treatment supplies).

The risk factors for type 1 and type 2 diabetes are different. People at increased risk of type 1 diabetes include:

- Children and young adults rather than older adults
- Caucasians, particularly people of Swedish or Finnish background

Risk factors for type 2 diabetes include:

- Family history of diabetes.
- Personal history of gestational diabetes or pre-diabetes.
- Age over forty-five.
- Obesity. 80 percent of persons diagnosed with type 2 diabetes are overweight.
- Ethnicity. African Americans, Hispanics, Native Americans, Pacific Islanders, and some Asian Americans are at greater risk than

Prior to the early 1920s, type 1 diabetes was not only incurable but fatal within a relatively short period of time. Although doctors were aware that the disease was caused by the lack of insulin secreted by the pancreas, early attempts to treat diabetics by feeding them fresh pancreas had failed. Frederick Banting (1891–1941), a Canadian doctor, worked together with a medical student named Charles Best at the University of Toronto to see whether they could successfully extract insulin from the pancreas and use it to treat diabetics. The two men first tested the extracted insulin on diabetic dogs. Their first human patient was a man named Leonard Thompson, who was close to death from diabetes. The success of Thompson's treatment led to Banting's winning the Nobel Prize in physiology or medicine in 1923.

Caucasians. Native Americans have the highest rates of type 2 diabetes of all ethnic groups in the United States.

- High blood pressure.
- High blood cholesterol levels.
- Not getting enough exercise. Exercise helps the body use blood sugar more efficiently.

# **Causes and Symptoms**

The basic cause of type 1 diabetes is an insufficient supply of insulin from the pancreas caused by an autoimmune disorder. What triggers the immune system's attack on the patient's pancreas, however, is not yet known. In type 2 diabetes, the patient's pancreas is secreting insulin but the body is not using the hormone efficiently—a condition known as insulin resistance. In type 2 diabetes, there are increased levels of insulin in the blood as well as sugar.

The central symptoms of both type 1 and type 2 diabetes are:

- Increased thirst
- Increased urination
- Increased appetite
- Fatigue

#### Diagnosis

The American Diabetes Association recommends that all adults over forty-five be screened for type 2 diabetes at least every three years, and that people with several risk factors be screened yearly.

Diabetes is diagnosed by the results of blood tests. There are several different types of blood tests that may be used.

- Fingerstick. This type of test is often used as a screener in the doctor's office. The doctor pricks the patient's fingertip with a needle and touches a test strip attached to a small handheld glucometer to the drop of blood, which provides a blood sugar reading within a few seconds. This method is used by diabetic patients at home to monitor their blood sugar levels.
- Fasting blood glucose test. The patient has blood drawn first thing in the morning after having eaten nothing since midnight of the previous night. A score over 126 milligrams per deciliter (mg/dL) indicates possible diabetes.

- Glucose tolerance tests. In this test, the patient has blood drawn twice, the first time after fasting for eight hours and the second time two hours later, after drinking a very sweet drink. A score over 200 mg/dL suggests diabetes.
- Hemoglobin A1c test. This test is used to monitor blood sugar control in people known to have diabetes. It measures the amount of blood sugar attached to the hemoglobin in red blood cells. If more than 7 percent of the hemoglobin has excess sugar attached to it, the person needs to get better control over their blood sugar levels.

# Treatment

There is no cure for diabetes. Treatment is based on a combination of diet and weight control; physical exercise; injected insulin or oral medications to reduce blood sugar levels; and home monitoring of blood glucose levels. The goal of treatment is to control blood sugar levels and to lower the patient's risk of blindness, frequent infections, heart disease, and other complications of diabetes:

- Diet: Patients with type 2 diabetes usually need to lower their weight. In addition, they must learn to eat roughly the same size meals at the same times each day so that their insulin doses will not lower their blood sugar level too quickly or too far.
- Exercise: Exercise reduces the patient's risk of leg ulcers, stroke, heart disease, kidney failure, and other complications of diabetes.
- Insulin and oral medications: Patients with type 1 diabetes must use injected insulin. Type 2 diabetics can be treated with oral medications that increase the body's sensitivity to insulin, stimulate the pancreas to produce more insulin, or decrease the amount of glucose produced by the liver. They may need insulin injections in addition to these oral medications.
- Home monitoring. Patients with either type of diabetes must check their blood sugar levels four times a day—before each meal and at bedtime—and keep a careful record of the test results.

# Prognosis

The prognosis of diabetes depends on the type and the patient's willingness to take responsibility for their health. Patients who do not take their insulin or other medications regularly or fail to monitor their blood sugar at home run the risk of severe complications.

According to the American Diabetes Association, people with diabetes are at risk for the following complications:

- Diabetics are two to four times more likely to develop heart disease than those without diabetes.
- Between 12,000 and 24,000 diabetics lose their eyesight each year.
- About 48,000 diabetics develop end-stage kidney disease each year.
- Around 71,000 amputations of legs and feet are performed each year in people with diabetes.
- Thirty-five percent of people with diabetes have periodontal disease.
- About 60–70 percent of diabetics have some degree of damage to the nervous system.

# Prevention

There is no way known to prevent type 1 diabetes, since its triggers are not yet understood.

People can prevent or lower their risk of type 2 diabetes by a combination of lifestyle changes and self-care:

- Keeping one's weight at a healthy level and eating a low-fat, high-fiber diet.
- Not smoking.
- Getting regular exercise, at least 30 minutes of walking or other activity five days a week.
- Keeping alcohol intake low.
- Getting regular checkups for blood pressure and blood cholesterol levels, and taking any medications prescribed by the doctor.
- If over forty-five, getting screened regularly for type 2 diabetes.

# The Future

Diabetes is expected to be a growing public health problem in the developed countries in the future. One reason is the aging of the population; the highest rates of diabetes in the United States are in people over 60. Another reason is the rise in obesity among children as well as adults.

# WORDS TO KNOW

**Endocrinologist:** A doctor who specializes in disorders of the pancreas and other glands.

Gestational: Pertaining to pregnancy.

**Glucometer:** A small blood testing device that can be used to screen for diabetes or used at home to monitor blood sugar levels.

**Insulin:** A hormone secreted by the pancreas that causes the cells in the liver, muscle and fatty tissues of the body to use the glucose carried in the bloodstream after a meal.

**Pancreas:** A small organ that lies between the stomach and the liver and secretes insulin.

The Centers for Disease Control and Prevention (CDC) predicts that one in three Americans born in the year 2000 will eventually develop type 2 diabetes. By 2050, the CDC expects the rate of diabetes in the United States to increase by 165 percent over the figures for 2006.

**SEE ALSO** Childhood obesity; Gangrene; Heart attack; Hypertension; Hypoglycemia; Obesity; Periodontal disease; Stroke

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# Down Syndrome

# Definition

Down syndrome, or trisomy 21, is a genetic disorder caused by the presence of an extra copy of chromosome 21 or by a portion of chromosome 21 translocated (attached) to another chromosome in one of the affected child's parents.

# Description

Children with Down syndrome have some degree of mental retardation (average IQ scores are 35–70) as well as characteristic facial features that include a head that is smaller than average, upward-slanting eyes, and a flattened nose. The hands are short and broad with short fingers, and they often have a single crease across the palm. Another characteristic feature of Down syndrome is hypotonia, which is the medical term for poor muscle tone. Children with Down syndrome often need extensive physical therapy in order to learn to walk and move normally. In addition, normal growth is slowed; most of these children never reach full adult height.

Babies with Down syndrome are often born with severe heart defects or blockages of the esophagus and small intestine. These conditions may require surgery shortly after birth. These children are also at increased risk of childhood leukemia.

#### Also Known As Trisomy 21

#### Cause

An extra copy of human chromosome 21

#### Symptoms

Small head; distinctive facial features; slow physical, mental, and social development

#### Duration

Lifelong



Young boy with Down syndrome. SHUTTERSTOCK.

Adolescents and adults with Down syndrome are more likely than other people to develop health problems that include frequent infections, cataracts, gastrointestinal reflux disease, hearing problems, sleep apnea, dislocated hips, and hypothyroidism.

# **Demographics**

Down syndrome occurs in about one in every 800 live births in the United States, or about 6,000 children per year. These babies, however, represent only about a quarter of those conceived with trisomy 21. The condition is linked to so many heart defects and other problems that affect survival before birth that about 75 percent of fetuses conceived with Down syndrome are miscarried.

Down syndrome occurs with equal frequency in all races and ethnic groups, as far as is known. Boys are slightly more likely to be affected than girls.

# **Causes and Symptoms**

Down syndrome results from genetic errors during the formation of germ cells (eggs and sperm) or during cell division shortly after the egg is fertilized by the sperm. The most common form of Down syndrome, responsible for about 95 percent of cases, occurs when an egg or sperm

# The Children's Advocate

John Langdon Haydon Down (1828-1896) was the British doctor who pioneered the care and education of children with the syndrome that now bears his name. He published the first scientific description of the condition in 1866. Down had originally hoped to become a chemist and studied under Michael Faraday, one of the most gifted scientists of his time, but then decided to enter medical school in 1853. Down was regarded as exceptionally talented; his professors predicted a brilliant future for him in medicine and were stunned when he turned down a prestigious position at the London Hospital to become superintendent of an asylum for mentally retarded children in Surrey in 1858. At that time, the field of mental retardation was considered unworthy of serious interest or concern, and affected children were regarded as beyond help.

For the next ten years Down worked at the Earlswood Asylum, turning it into a model institution for the care of mentally ill as well as retarded children and adolescents. In 1868 he founded a school for the education of children with trisomy 21, which was then termed mongolism. Down believed that these children could indeed learn and contribute to society. He also advocated for higher education for women, arguing against the widespread belief that allowing females to study at the university level would make them more likely to have retarded children.

Two of Down's sons became doctors and continued his work at the school he founded. Although the genetic cause of trisomy 21 was not known in Down's day—he himself attributed it to tuberculosis in the children's parents—he was an important advocate for those affected by the syndrome. The cause of Down syndrome was finally identified in 1959 by Jérôme Lejeune (1926–1994), a French pediatrician and geneticist.

carrying two copies of chromosome 21 is involved in conception. The reason for the extra copy in the abnormal germ cell is a genetic error called nondisjunction. During the normal process of germ cell formation, the paired chromosomes in the cell divide so that each daughter cell has only one member of the pair. In nondisjunction, one daughter cell gets both members of the chromosome pair and the other cell has none. If a germ cell carrying two copies of chromosome 21 is fertilized by a normal germ cell from the other parent, the child will have three copies of chromosome 21. This genetic error is called a full trisomy 21.

Some children with Down syndrome have some body cells with the extra copy of chromosome 21 and some body cells without the extra copy. This condition is called mosaic trisomy 21. It is thought to result from random errors in cell division during the early stages of fetal development. Mosaic trisomy 21 accounts for about 2 percent of children with Down syndrome.

About 3 percent of cases of Down syndrome occur in families. A part of chromosome 21 may become attached to chromosome 14 either before or at the moment of conception. This type of genetic error is called a balanced translocation because there is no extra material from chromosome 21. A person with this type of translocation looks normal and develops normally; however, he or she has an increased risk of having a child with full trisomy 21.

The mother's age is a risk factor for Down syndrome, rising from one chance in 1,562 in mothers age twenty-four or younger to one in nineteen in mothers over age forty-five. Recent studies indicate that the father's age is also a factor; men forty-two years and older are at increased risk of having a child with trisomy 21.

In addition to the physical features mentioned earlier, other indications of Down syndrome in a newborn include:

- An additional skin fold at the inner corner of the eye
- A short neck
- White spots on the iris of the eye known as Brushfield spots
- A round face
- Ears that are smaller than normal
- A flattened area at the back of the head
- · Missing teeth or delayed development of teeth
- Protruding tongue and a tendency to breathe through the mouth
- An extra-wide space between the big toe and second toe

## Diagnosis

Most babies with Down syndrome are diagnosed at birth on the basis of their physical features. The diagnosis can be confirmed by a blood test and karyotype (an analysis of a person's chromosomes).

The diagnosis can also be made before birth. There are two types of prenatal tests: screeners and diagnostic tests. Screeners only estimate the baby's risk of having Down syndrome. Given between the fifteenth and twentieth weeks of pregnancy, screening tests include a blood test and an ultrasound imaging test. These tests are only about 60 percent accurate, however. Diagnostic tests that are about 98 percent accurate include chorionic villus sampling (CVS), done between the ninth and fourteenth weeks of pregnancy; amniocentesis, which can be done at the same time that the screeners can be given; and testing blood samples taken from the baby's umbilical cord, done after the eighteenth week of pregnancy.

## Treatment

The treatment of children with trisomy 21 is highly individualized. Babies with heart defects or obstructions in the esophagus and digestive tract usually need immediate surgery. Older children require periodic checkups for cataracts, hearing loss, and thyroid problems. Some children need special medications and diuretics for heart problems, and most need to be monitored for frequent infections, particularly ear infections and pneumonia.

At one time children with Down syndrome were either institutionalized or put in special education programs apart from other children. By the early 2000s, however, the emphasis in treatment was to give these children as many opportunities as possible to go to school with other children in their age group and participate in sports, group activities, and other aspects of social life during the growing years.

## Prognosis

People with Down syndrome have a shortened life expectancy and a high risk of early Alzheimer disease, often showing a noticeable loss of mental function by age forty. About 85 percent of babies born with trisomy 21 survive the first year of life, but only 50 percent will live to reach age fifty, according to data available in 2008. This is a great improvement, however, as the life expectancy of a person with Down syndrome was only twenty-five years as recently as 1980.

Congenital heart disorders are one reason for the present high mortality rate, as are vulnerability to infections, a high rate of disorders of the digestive tract, and premature aging. Children with Down syndrome are also more likely to develop leukemia than other children.

It is not possible to tell at birth whether a baby with Down syndrome will be severely retarded or will have low-normal intelligence. Individualized assessment of the child is critical to providing opportunities for full development. In general, children with mosaic Down syndrome have higher IQ scores than children with full trisomy 21. There are many adults with the syndrome who are able to hold jobs and live independently; some have become successful artists, actors, and singers.

# WORDS TO KNOW

**Chorionic villus sampling (CVS):** A prenatal test that involves taking a small sample of the placenta, the organ that forms inside the uterus during pregnancy and supplies the baby with oxygen and nutrients carried by the blood.

Congenital: Present at birth.

- **Germ cell:** A cell involved in reproduction. In humans the germ cells are the sperm (male) and egg (female). Unlike other cells in the body, germ cells contain only half the standard number of chromosomes.
- **Hypotonia:** The medical term for poor muscle tone.
- **Karyotype:** A photomicrograph of the chromosomes in a single human cell. Making a karyotype is one way to test for genetic disorders.
- **Mosaicism:** A condition in which a person has some body cells containing an abnormal number of chromosomes and other cells containing the

normal number. Mosaicism results from random errors during the process of cell division that follows conception.

- **Nondisjunction:** A genetic error in which one or more pairs of chromosomes fail to separate during the formation of germ cells, with the result that both chromosomes are carried to one daughter cell and none to the other. If an egg or sperm with a paired set of chromosomes is involved in the conception of a child, the child will have three chromosomes in its genetic makeup, two from one parent and one from the other.
- **Translocation:** A genetic error in which a part of one chromosome becomes attached to another chromosome during cell division.
- **Trisomy:** A type of genetic disorder in which a cell contains three copies of a particular chromosome instead of the normal two. Down syndrome is one of several trisomies.

# Prevention

Since most cases of Down syndrome are caused by a spontaneous genetic mutation rather than an inherited genetic defect, there is no completely effective way to prevent trisomy 21. Adults who are concerned that they may have a balanced translocation of chromosome 21 can choose to have a karyotype to see whether their chromosomes are in fact abnormal. Pregnant women over thirty-five should have tests during the first trimester (three-month period) of pregnancy to screen for the syndrome.

# The Future

It is possible that the increasing numbers of women having children in their thirties or forties will lead to an increase in the number of children born with Down syndrome. However, as of the early 2000s, about 90 percent of women whose fetuses were diagnosed with the syndrome chose to end their pregnancies before childbirth. There has been growing concern as to whether such women are being pressured to make this choice. Parents have formed advocacy groups to defend their choice to have their children in spite of the prenatal diagnosis, and disability rights groups have also spoken on their behalf. The first World Down Syndrome Day was held in Singapore in 2006 to raise awareness of the many positive contributions of people with trisomy 21. Sarah Palin, 2008 Republican Vice Presidential candidate, has a son with Down syndrome and raised awareness about children with special needs during her campaign.

**SEE ALSO** Alzheimer disease; Congenital heart disease; Edwards syndrome; Gastroesophageal reflux disease; Hypothyroidism; Leukemia; Patau syndrome; Triple X syndrome

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# Dwarfism

See Achondroplasia.

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# Dyslexia

# Definition

Dyslexia is a learning disability related to reading and writing that is neurological in origin. It is the most common learning disorder among children of school age.

# Description

Dyslexia is not a single condition but rather a cluster of symptoms related to reading difficulties and other language skills. Children with dyslexia usually have problems pronouncing words and spelling them correctly as well as difficulties with writing words and sentences. Most people with dyslexia have trouble identifying the separate speech sounds within a word or learning how letters represent those sounds. This particular language skill is known as decoding. Although there is a popular belief that dyslexia is a visual problem that involves reading letters or words backward or upside down, this belief is not true. Reversals of letters or words are also not a defining characteristic of dyslexia.

Dyslexia is not associated with mental retardation; it can affect children at all levels of intelligence. Most children with dyslexia are of normal intelligence; some are highly gifted, and many excel in fields that do not require high-level language skills, including the visual arts, computer science, design, drama, electronics, math, mechanics, music, physics, dance, and athletic competition.

Dyslexia does not affect all children equally. Some children diagnosed with the condition are able to master the basics of reading and Also Known As Reading disorder, reading disability

#### Cause

Differences in the way the language areas in the brain develop and function

#### Symptoms

Difficulties in word recognition, reading, and writing

Duration Lifelong Dyslexic girl writing in notebook, showing the transposition of letters in words. © WILL AND DENI MCINTYRE/PHOTO RESEARCHERS, INC.

> spelling, particularly if they have competent and dedicated teachers. They may, however, run into difficulty when they reach the upper grades and are confronted with the need for more complex language skills that include a knowledge of grammar, the ability to read and remember textbook material, and the ability to write longer compositions.

> Some children with dyslexia have problems with understanding spoken language even though they have teachers and parents with good spoken language skills. These children may have trouble expressing themselves or may misunderstand what others say. Problems with spoken language place this group of dyslexics at a severe disadvantage not only in school and later on in the workplace but also in their relationships with other people.

# **Demographics**

Dyslexia is thought to affect about five out of every 100 school-age children in the United States, with another 5–10 percent having difficulty with reading. Some researchers think that as many as 20 percent of American adults have some of the symptoms of dyslexia, including slow or inaccurate reading, problems with spelling and writing, or mixing up words that sound similar.

At one time it was thought that boys were more likely to be dyslexic than girls; however, researchers now think that boys and girls are equally likely to have the disorder.

# The Fonz Becomes a Writer

Henry Winkler (1945–), the actor who played the Fonz in the long-running television show *Happy Days*, had a miserable childhood because of undiagnosed dyslexia. He recalls that lunch was the only subject he was good at doing. Even his parents thought he was dumb and lazy. Winkler eventually finished his education, including a master's degree from the Yale School of Drama, and went on to become a well-regarded actor but still has painful memories of his early years. He was finally diagnosed with dyslexia at the age of thirtyone, when his stepson was also diagnosed with the disorder.

Winkler decided to make a career change and help children who have trouble with language skills. He has produced a series of books together with a writer named Lin Oliver about a boy named Hank Zipzer, "The World's Greatest Underachiever." The books follow Hank's adventures at Winkler's old school, P.S. 87 in New York City. Winkler told an interviewer in 2007 that he tries to make the books as goodhumored as possible to encourage young readers. He was delighted by a letter he received recently



Henry Winkler as Fonzie from the television show "Happy Days." SILVER SCREEN COLLECTION/HULTON ARCHIVE/ GETTY IMAGES.

from a boy in Montana who said, "I laughed so hard, my funny bone fell out of my body."

# **Causes and Symptoms**

The causes of dyslexia are not completely understood but are thought to be related to differences in the structure and functioning of the brain. Dyslexics appear to process information in a different part of the brain than nondyslexics. Although the condition has not yet been conclusively traced to specific genes, it is known to run in families. In 2005, researchers identified a region on chromosome 6 as possibly related to dyslexia, but this finding has not yet been verified.

One surprising finding, reported by a team of researchers at Hong Kong University in 2008, is that the part of the brain affected by dyslexia appears to differ according to the child's primary language. The researchers used magnetic resonance imaging (MRI) to compare a group of children whose first language is English with a second group raised to speak Chinese. The scientists found that the English speakers use a different part of the brain when reading from that used by the Chinese students. The difference is apparently related to the fact that English is an alphabetic language whereas Chinese uses symbols to represent words.

In addition to having trouble associating letters with sounds and forming memories for words, children with dyslexia may have some of the following problems with learning:

- · Learning to speak.
- Organizing thoughts and ideas into clear written and spoken language.
- Memorizing number facts, such as the multiplication tables.
- Reading quickly enough to understand what is being read. Some children with dyslexia read so slowly that they cannot remember the beginning of a sentence by the time they reach the end of it, particularly if it is a long and complicated sentence.
- Making their way through longer reading assignments.
- Spelling words correctly.
- Trouble with making rhymes.
- Learning foreign languages, which involves a basic understanding of grammar and the parts of speech.
- Performing mathematical calculations correctly.

#### Diagnosis

The diagnosis of dyslexia can be a complicated process that usually involves a psychologist and an education expert as well as the child's family doctor. The first step is to rule out any disorders of vision or hearing that could interfere with learning. The child may also be referred to a neurologist (a specialist in disorders of the nervous system) for further evaluation to make sure that the child does not have a brain tumor or other physical disease of the brain.

The next step is usually intelligence testing and an evaluation of the child's reading and speaking skills by a qualified expert. This type of evaluation involves testing the child's short-term memory or asking the child to read nonsense words as a test of his or her ability to link letters and sounds. In addition, the child may be evaluated psychologically to see whether depression, anxiety, or social problems are causing the learning difficulty.

## Treatment

There is no medical or surgical cure for dyslexia; however, dyslexic individuals can learn to read and write with appropriate education or treatment. Treatment for dyslexia is highly individualized. After the child's specific difficulties in reading and understanding language have been analyzed by an expert, a treatment program is drawn up tailored to his or her needs. Most children with dyslexia benefit from a multisensory approach to language. A multisensory approach is one that involves several senses (seeing, hearing, touching) all at the same time. Teachers who use this approach teach children to link the sounds of the letters with the written symbol. Children also link the sound and symbol with how it feels to form the letter or letters. Another way to use the multisensory approach is to have the child listen to an audiotape while tracing the shapes of letters with the fingers and the words spoken.

Children with severe dyslexia may require tutoring on a one-to-one basis or in small group sessions several times a week. Whereas a child with normal language skills may need sixty to ninety hours to master a specific set of tasks involving reading, children with dyslexia may need between eighty and 100 hours to make the same progress. In general, the earlier a child is diagnosed and special education programs are started, the greater the likelihood that he or she will learn to read well enough to succeed in school. Some children with severe dyslexia, however, may never learn to read or write well and are usually helped by training for occupations or career paths that do not require strong language skills.

Children with dyslexia often need and benefit from psychotherapy because of their struggles with low self-esteem. Many come to feel that they are stupid or less capable than they really are; they are likely to drop out of school if they are not diagnosed and treated early.

#### Prognosis

The prognosis of dyslexia is difficult to predict because an individual child's outcome depends on a number of factors: the severity of the child's language difficulties; the stage at which the dyslexia is diagnosed; the supportiveness of the child's family; the presence of other health problems; and the quality of supplemental teaching and tutoring that the child receives. Many people with dyslexia, however, do achieve personal and professional success; some go on to complete advanced degrees while others do well in such fields as business and the performing arts.

# WORDS TO KNOW

**Decoding:** In education, the ability to associate letters of the alphabet with sounds.

**Neurologist:** A doctor who specializes in diagnosing and treating disorders of the nervous system.

# Prevention

There is no known way to prevent dyslexia.

## The Future

Research in dyslexia is focused not only on experimentation with new types of educational techniques but also on research into the genetic factors involved in dyslexia. In addition, ongoing work with such tools as magnetic resonance imaging and positron emission tomography (PET) may yield new insights into the structural differences between the brains of children with dyslexia and the brains of children with normal language skills.

Still another area of research is the relationship between specific languages and children's difficulty in learning to read or speak. Some scientists have suggested, for example, that English-speaking children have a harder time learning to read than Italian-speaking children because English has many more irregular spellings and pronunciations than Italian.

**SEE ALSO** Developmental disability

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# E



**Also Known As** Otitis media, otitis externa,

swimmer's ear

**Cause** Bacterial, viral, or fungal infection

#### Symptoms

Pain or fullness in the ear, hearing loss, loss of balance

#### Duration

About a week (outer ear); one to two weeks (middle ear)

# E. Coli See Food poisoning.



# Ear Infection

## Definition

Ear infections are caused by either a bacterial or fungal infection of the outer ear or a viral or bacterial infection of the middle ear. The outer ear is the visible part of the ear plus the ear canal, a small passage that conducts sound waves from the outside to the middle ear.

The middle ear is a group of structures that include the eardrum and three small bones called ossicles that convey sound energy from the ear canal to the structures of the inner ear. The middle ear is connected to the upper throat by a passageway called the Eustachian tube, which has two major functions: to equalize air pressure between the middle ear and the air outside the body and to drain fluid or mucus from the middle ear into the throat.

# Description

Infection of the outer ear, or otitis externa, is a skin infection caused by bacteria or fungi that get into the skin of the ear canal through a scratch or other small break in the skin. The skin lining the ear canal is very thin and easily damaged. If a person goes swimming, takes a shower, or is

#### Ear Infection

Inflamed eardrum that signals an ear infection. © MEDICAL-ON-LINE / ALAMY.



exposed to hot, humid weather, bacteria or fungi in the ear canal can rapidly multiply and cause an inflammation of the skin. Because the skin of the ear canal is closely attached to the underlying bone, the inflammation can cause severe pain and swelling that may be sudden. The swelling of the skin of the canal may lead to temporary hearing loss.

Infection of the middle ear, or otitis media, is often a complication of the common cold. It begins when cold viruses (or sometimes bacteria) enter the Eustachian tube from the upper throat and produce inflammation and swelling in the tube. Fluid builds up behind the eardrum when the Eustachian tubes are blocked by swelling; this is called an effusion. The Eustachian tubes can also be blocked by swollen adenoids, pieces of tissue that are part of the immune system and lie at the very back of the nasal passages. If the adenoids become infected, they can swell and block the Eustachian tubes.

Doctors usually distinguish between otitis media with effusion (OME) and acute otitis media. In OME, the fluid that builds up behind the eardrum is not itself infected; in acute otitis media, the collection of fluid itself has become infected by bacteria. The difference is important because it affects the treatment of the earache.

# Demographics

Infection of the outer ear is common. In the United States, it is more common in the summer months and in the warmer and more humid parts of the country and is more likely to affect adolescents and young adults than very young children.

Infection of the middle ear, however, is much more common in young children than in older children or adults. There are two reasons for this. First, the immune systems of young children are less well developed than those of older children; second, the Eustachian tubes in young children enter the upper throat at a lower angle than in older humans. This difference makes it easier for disease organisms to stay in the tubes and cause inflammation and swelling rather than being carried downward into the throat.

Infections of the middle ear are very common in children between six months and three years of age. According to the National Institutes of Health (NIH), 50 percent of all children in the United States have at least one episode of otitis media by the time they are a year old, and 80 percent have an episode by three years of age. The costs of treating these infections and their complications come to \$4 billion each year.

Otitis media is more common in the fall and winter months in the United States. It is somewhat more common in boys than in girls and is more common in Native Americans than in children of other racial groups. The reasons for these differences are not known.

Factors that increase a child's risk of middle ear infections include:

- Heredity. Repeated infections of the middle ear are known to run in some families, although no specific genes have been linked to otitis media.
- Having a cleft palate.
- Day care. Children in day care settings are exposed to common colds and other upper respiratory infections that can lead to otitis media.
- Exposure to tobacco smoke or air pollution.
- Feeding position. Babies who are fed from a bottle while lying down are at greater risk of ear infections than those who are held upright.
- Family history of frequent ear infections.
- Use of a pacifier.
- History of allergies.

#### **Causes and Symptoms**

Ear infections are caused by disease organisms causing tissue inflammation and fluid buildup in the skin of the outer ear or the structures of the middle ear. The symptoms of otitis externa may include:

- Sudden onset of pain
- Intense pain when the outer ear is pulled or moved
- Itching
- Swelling of the outer ear or nearby lymph nodes in the neck
- Feeling of fullness in the ear
- Temporary loss of hearing or feeling that sounds are muffled
- Pus draining from the ear
- Red or flaky skin on the outside of the ear

The symptoms of otitis media may include:

- Intense crying in very young children
- Tugging or pulling at the ear
- Fever
- Irritability and headaches
- Trouble sleeping or poor feeding
- Nausea and vomiting (in small infants)
- Hearing loss
- Ringing or buzzing sounds in the ear
- Fluid draining from the ear (This symptom usually indicates that the eardrum has ruptured.)

If the child has otitis media with effusion, there may be a slight hearing loss or no symptoms at all.

#### Diagnosis

The diagnosis of an ear infection is based on a combination of the patient's age, history, and a physical examination in the doctor's office. Otitis externa can usually be diagnosed by simple movement of the outer ear, which will typically produce intense pain. When the doctor looks into the ear with an otoscope, the ear canal will look red and swollen, and there may be pus present. The doctor may take a sample of the pus or fluid and send it to a laboratory for culture.

In the case of otitis media, the doctor will use a pneumatic otoscope to examine the child's ear. This specialized otoscope allows the doctor to puff a small amount of air into the middle ear to see whether there is fluid behind the eardrum. If fluid is present, the eardrum will not move. Another test known as tympanometry may also be done to measure the movement of the eardrum. In tympanometry, a small plug is inserted into the outer ear and air is blown into the ear canal to evaluate the movement of the eardrum.

If there is evidence of hearing loss, the child may be referred to an audiologist for hearing tests.

## Treatment

Treatment of an infection of the outer ear may involve one or more of the following:

- Cleaning the outer ear with a cotton swab or suction device to remove flaky skin and pus or other fluid.
- Antibiotic ear drops to fight infection. If the ear canal is swollen shut, the doctor may insert a wick that will allow the drops to penetrate the full length of the ear canal.
- Aspirin or ibuprofen to relieve pain and reduce inflammation.
- Ear drops containing a steroid medication to reduce itching and tissue swelling.

The patient will be told to avoid swimming or scuba diving until the infection is cleared and to keep water out of the ears while bathing or showering.

Treatment of otitis media depends in part on whether the patient has otitis media with effusion (OME) or a bacterial infection. If the swelling of the Eustachian tube and the fluid buildup are caused by a virus, antibiotics will not help. About 80 percent of children with otitis media do not have a bacterial infection and will recover without antibiotics.

The American Academy of Pediatrics (AAP) recommends a waitand-see approach for the first two to three days to see whether the infection will improve without antibiotics. Parents can give the child nonaspirin pain relievers to relieve fever, apply warm washcloths to the outer ear, or use anesthetic ear drops for pain. Antibiotics are usually prescribed, however, for babies younger than six months; older children who have had two or more ear infections within a month; children in severe pain; or children with a fever of 102°F (38.9°C) or higher.

The doctor may recommend surgical treatment if the child has recurrent infections of the middle ear or if the infections are not cleared by antibiotics. In this type of surgery, a small drainage tube is inserted through the eardrum to drain fluid and to equalize the pressure between the middle ear and outer ear. The tubes usually fall out on their own as the child grows. If the child's Eustachian tubes are blocked by swollen adenoids, the doctor may recommend surgical removal of the adenoids.

## Prognosis

Infections of the outer ear usually clear up completely in about a week without long-term complications. In some cases, however, people develop a chronic infection of the outer ear that extends to inflammation of the surrounding skin. A few people, most often those with diabetes or a weakened immune system, may develop a severe infection of the bone and cartilage near the outer ear that can cause severe pain and spread to the brain. This rare but potentially life-threatening complication requires treatment with intravenous antibiotics and sometimes surgery.

Most cases of otitis media improve within two to three days and clear up completely in a week or two without complications. If fluid remains behind the eardrum for long periods of time, however, it may eventually cause hearing loss. Another possible complication of recurrent or untreated otitis media is the spread of infection into air cells called mastoids in the bones around the base of the skull, a condition known as mastoiditis.

## Prevention

Infections of the outer ear can be prevented by using ear plugs when swimming, avoiding swimming in polluted water, drying the ears after swimming or showering, and avoiding the use of foreign objects to clean wax out of the ears. It is very easy to damage the skin of the ear canal in this way.

Infections of the middle ear can be prevented by keeping a child away from children with colds or upper respiratory infections; by not exposing the child to tobacco smoke; by feeding the child in an upright position; and by breastfeeding the child for the first six months of life. Some doctors also recommend giving the child Prevnar, a vaccine that protects against pneumonia and appears to reduce the risk of otitis media as well.

# WORDS TO KNOW

**Effusion:** The medical term for an abnormal collection of fluid in a body cavity.

**Eustachian tube:** The passageway that connects the middle ear with the upper throat.

Otitis: The medical term for inflammation of the ear.

# The Future

Ear infections are likely to continue being common health problems in children and adolescents. Researchers are comparing the effectiveness of tube placement versus removal of the adenoids in treating otitis media. They are also studying the effectiveness of the pneumonia vaccine in preventing infections of the middle ear.

SEE ALSO Cleft lip and palate; Common cold; Smoking

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# **m** Eating Disorders

Eating disorders are a group of mental disorders related to abnormal eating patterns. Patients diagnosed with eating disorders feel a need to lose weight, sometimes cutting back on eating to the point of malnutrition and starvation. They may also exercise for hours or force themselves to vomit in order to keep their weight as low as possible. Some people with eating disorders alternate between eating as little food as possible and episodes of binging. Binging refers to eating large quantities of food in a brief period of time and being unable to control the eating. This is often followed by vomiting.

Eating disorders are considered mental disorders because they involve distortions of reality. Specifically, people with eating disorders have a distorted image of what their bodies really look like. They are also often perfectionists—unwilling to accept their limitations and often feeling powerless. Controlling food intake becomes a way of trying to control all aspects of their lives.

Eating disorders were extremely rare until the 1970s. Many doctors think that society's emphasis on physical attractiveness, particularly on thinness as essential to beauty, is a major reason for the rapid spread of eating disorders in developed countries over the last forty years. Although the majority of patients with eating disorders are women, between 10 and 12 percent are male.

SEE ALSO Anorexia; Bulimia



# Ebola and Marburg Hemorrhagic Fevers

# Definition

Ebola and Marburg fevers are two forms of viral hemorrhagic fever (VHF), a devastating infection with a very high mortality rate. Both are considered emerging infectious diseases because they were unknown before the mid-1960s. Both are also considered zoonoses because humans are thought to get them from contact with the tissues or body fluids of infected animals. Once the virus infects a human, the infections can spread from person to person.

Both Ebola and Marburg fever are caused by an unusual type of virus called a filovirus. Filoviruses look like long looped threads when viewed under an electron microscope. There are four known strains of Ebola virus but only one strain of Marburg virus.

# Description

Marburg virus was the first identified filovirus, named for the city in Germany where seven laboratory staff members died after working with green monkeys imported from Uganda. The monkeys were being used to develop polio vaccines. Two doctors in Marburg became infected by contact with blood drawn from the first group of patients. Later cases of Marburg virus infection resulted from tourists visiting caves in Africa where infected bats are known to roost, or from direct contact with infected humans. An outbreak in the Congo from 1998 to 2000 was spread by gold miners.

Ebola virus is named for the river valley in the Democratic Republic of the Congo (then Zaire) where the first outbreak in humans occurred in 1976. Three of the four subtypes of Ebola virus are known to cause disease in humans. The fourth, known as Ebola Reston, was identified at the U.S. Army research laboratories (USAMRIID) in Reston, Virginia, in 1989. It causes disease in monkeys but not in humans. Researchers think that tropical fruit bats are the reservoir of Ebola as well as the Marburg viruses. The bats can give the filoviruses to monkeys, humans, and other animals, but do not get sick themselves.

#### Also Known As

Ebola HF, Marburg HF, viral hemorrhagic fever, green monkey fever

# Cause

Filoviruses

#### Symptoms

Fever and flu-like symptoms followed by a rash, vomiting and diarrhea, and severe bleeding

#### Duration

One to three weeks of acute symptoms if the patient survives

#### Ebola and Marburg Hemorrhagic Fevers



Both Ebola and Marburg fevers have a high mortality rate—from 23–90 percent for Marburg fever and 50–90 percent for Ebola. Humans usually get infected by direct contact with the blood, other body fluids, or tissues of infected animals or other humans. Healthcare workers and family members preparing the dead for burial have been at highest risk. Viral hemorrhagic fevers typically begin suddenly with flu-like symptoms (fever, headache, muscle and joint pains) followed rapidly by a skin rash, violent nausea and vomiting, bloody diarrhea or bleeding from the nose and other parts of the

*Image of the Ebola virus.* © ALAMY/ ALAMY.

body, mental confusion, and depression. Death results from a loss of blood volume and massive organ failure. Not all viral hemorrhagic fevers are caused by filoviruses.

## Demographics

In the United States, there were no documented cases of Ebola or Marburg fever as of 2008. There have been four instances in 1989 and 1990 when several animal care workers in research laboratories tested positive for infection with the Ebola Reston strain of the virus. The workers did not become sick, however. The chief concern of public health doctors in Canada and the United States is the possibility that the Marburg and Ebola viruses could be used as agents of bioterrorism.

According to the World Health Organization (WHO), viral hemorrhagic fevers are equally deadly to men and women, and to members of all races. Children appear to be less likely to become infected, but that is most likely because they have less contact with sick adults than healthcare workers or adult caregivers.

The risk of someone living in North America being infected with a viral hemorrhagic fever is very low. The following groups of people are at increased risk, however:

- Those having jobs involving frequent travel to areas of the world with known outbreaks of Ebola or Marburg fever. These include the Democratic Republic of the Congo (formerly Zaire), Sudan, Angola, Gabon, and Côte d'Ivoire.
- Field biologists who travel to Africa to study fruit bats.

- Researchers who study monkeys or other primates imported from Africa.
- Doctors, nurses, and other caregivers who treat or care for people infected with Marburg or Ebola virus.

# **Causes and Symptoms**

Both Marburg and Ebola fever are caused by filoviruses that usually enter the body through direct contact with blood, urine, vomit, or other body fluids from infected animals or people. The viruses can also be spread by reusing contaminated needles or syringes. Some researchers think it may be possible for the viruses to spread through the air as well, as some monkeys appear to have become infected by breathing dust from bat droppings.

Once inside a human body, filoviruses replicate rapidly during an incubation period of three to nine days. They target the tissues that line the inner walls of blood vessels, causing tissue destruction, problems with blood clotting, and the hemorrhaging that is characteristic of these fevers. The liver is the internal organ that is most severely damaged by these diseases.

Patients usually feel sick quite suddenly. Viral hemorrhagic fevers can be difficult to diagnose at this point because the first symptoms are similar to those of malaria, yellow fever, typhoid fever, flu, or various bacterial infections. The patient has a high fever (102°F/39°C), sore throat, severe headache, nausea, dizziness, abdominal pain, muscle aches, and general weakness. In the next stage, the patient may have a reddish skin rash resembling measles; red eyes due to hemorrhaging of the tiny blood vessels in the eyes; vomiting blood; hemorrhaging blood from the nose, mouth, ears, or anus; severe diarrhea; and abnormally low blood pressure.

Flioviruses can destroy the patient's kidneys, lymph nodes, lungs, and spleen as well as the liver. Death occurs between seven and fourteen days after the first symptoms as a result of shock and organ failure.

# Diagnosis

A correct diagnosis of Ebola or Marburg fever requires the doctor to suspect that the patient has an illness more serious than typhoid or malaria. The patient's history—particularly a history of travel to Africa or an occupational history of working with animals—may be the most important clue.
The viruses that cause Ebola and Marburg fever can be detected in blood and tissue samples, but these must be carefully collected and sent to the Centers for Disease Control and Prevention (CDC) or another laboratory equipped to handle samples that are considered a level 4 biohazard (the highest possible rating). The best tests for detecting filoviruses are the enzyme-linked immunosorbent assay or ELISA test, and the reverse transcriptase polymerase chain reaction or [RT-]PCR test.

## Treatment

There is no specific treatment for viral hemorrhagic fevers. Patients must be taken to a hospital for supportive care, which consists of fluid replacements, replacing blood loss, monitoring the patient's blood pressure, and prescribing medications to treat any bacterial infections that may develop in addition to the viral infection.

Patients diagnosed with Ebola or Marburg fever must be kept in strict isolation from other patients, and the doctors and nurses who care for them must wear goggles, face shields, and gowns as well as rubber gloves at all times. Any object that has been in contact with the patient's body fluids must be completely disinfected with chlorine bleach.

### Prognosis

The prognosis for surviving Ebola virus is poor. The disease has a mortality rate as high as 90 percent. Patients who live for two weeks after symptoms appear have a chance of making a slow recovery that takes months.

Marburg hemorrhagic fever has a mortality rate between 23 and 90 percent. As with Ebola fever, survivors may take months to regain their strength. The after-effects of both diseases may include hair loss, eye problems, hepatitis, weakness, headaches, and (in men) inflammation of the testicles.

### Prevention

The best way to avoid infection with a filovirus is to avoid traveling to parts of the world with known outbreaks of viral hemorrhagic fever. People should also avoid all contact with the remains of dead monkeys or other primates in these areas.

In the event of an outbreak, those who die from Ebola or Marburg fever must be buried as quickly as possible by specially trained workers in order to lower the risk of the infection spreading farther.

## WORDS TO KNOW

**Bioterrorism:** The use of disease agents to frighten or attack civilians.

- **Emerging infectious disease (EID):** A disease that has become more widespread around the world in the last twenty years and is expected to become more common in the future.
- Filovirus: The category of viruses that includes Ebola and Marburg viruses.

Filoviruses look like long pieces of thread under a microscope.

- **Reservoir:** The term used by biologists for the natural host species of a disease organism. Bats are thought to be the reservoir of viral hemorrhagic fevers.
- **Zoonosis (plural, zoonoses):** A disease that animals can transmit to humans.

### The Future

Researchers are presently working on developing a vaccine for humans against Ebola and Marburg fever. Vaccines that are 99 percent effective in monkeys have already been developed. However, early trials of a vaccine for humans were not successful. As of 2008, a newer vaccine for use in humans had entered clinical trials and seemed to be effective. Further study is needed before it can be approved for use.

Some doctors are concerned that Ebola and Marburg viruses could be used as weapons of bioterrorism. The filoviruses are classified by the CDC as Category A agents (the highest category) along with smallpox and anthrax. Category A agents are disease agents that have high fatality rates and could be spread across a large population. There are, however, two factors that could limit the use of filoviruses by bioterrorists. One is that Ebola and Marburg viruses are not easily spread among humans through the air. There is only one known case of a person being infected by airborne droplets coughed up by an Ebola patient. The other limitation on the use of these viruses in bioterrorism is that they infect everyone in a small community very rapidly but then die out before reaching larger human groups. In any case, the possibility of these viruses being used by terrorists is a major motivation for speeding the development of safe and effective vaccines against them.

SEE ALSO Anthrax; Smallpox; West Nile virus infection

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Also Known As Atopic dermatitis, AD

#### Cause

Possible immune system overreaction

### Symptoms

Thickened, scaly, or discolored skin, intense itching, open sores

### Duration

May clear up after childhood or be lifelong



# Eczema

# Definition

Eczema is a noncontagious skin disease characterized by extremely itchy inflamed skin that often becomes cracked or weepy after scratching. It is grouped together with asthma and hay fever as an atopic disease. Atopic diseases are allergic conditions in which the affected parts of the body are not in direct contact with the allergen. In addition, these diseases are often inherited.



*Eczema rash on a patient's leg.* © PHOTOTAKE INC. / ALAMY.

# Description

People with eczema develop an itching, scaling, and swelling rash on the skin that may form oozing open sores or yellow and red pimples. The rash is most likely to affect the skin of the hands and feet, the arms, the area behind the knees, and the ankles, wrists, face, neck, and upper chest. Some people develop eczema on their eyelids or the skin around the eyes. Scratching in this facial area may eventually cause the eyebrows and eye-lashes to fall out. Other people may develop an extra fold of skin under the eyes from rubbing or scratching.

The most troublesome symptom of eczema for most patients is the intense itching that accompanies the disease. An officer of the National Eczema Association said in an interview, "It's like having poison oak or poison ivy twenty-four hours a day, seven days a week, forever." The itching in turn can lead to sleeping difficulties—children with severe eczema may lose as much as two hours of sleep per night—as well as self-consciousness in social situations and other psychological disturbances.

# **Demographics**

Eczema is more common among children than adults; 95 percent of cases occur in children five years or younger. Between 10 and 12 percent of American children are diagnosed with eczema. Of these children,

# Self-Care for Eczema

People who are affected by eczema can care for themselves at home by:

- Moisturizing the skin frequently, especially after bathing or showering. Most doctors recommend applying the moisturizer within three minutes after the bath. The bath or shower water should be comfortably cool rather than hot. Some people are helped by adding baking soda or Aveeno (an oatmeal product) to their bath water. Soap or bath gels should be mild and (if possible) unscented.
- Patting the skin dry rather than rubbing vigorously with a towel.
- Wearing clothing made of cotton and wearing several layers of it in the winter rather than using wool. Wool rugs and blankets should also be avoided as they can irritate the skin.
- Using a humidifier during heating season to keep the air in the house as moist as possible.
- Avoiding extremes of temperature whenever possible. Hot and humid summer weather can cause flare-ups in some patients because of increased sweating.
- Exercising at a moderate level during flare-ups. Intense physical activity can make flare-ups worse.
- Avoiding foods that are known to trigger flare-ups.
- Reducing emotional stress. Some people with eczema are helped by stress management techniques or starting a meditation practice.

30 percent will develop asthma and 35 percent will develop hay fever in later childhood.

The disease often goes into remission (quiet period without symptoms) in late childhood or adolescence and then flares up again in the early adult years. According to the American Academy of Dermatology (AAD), about half of children diagnosed with eczema will improve by the time they are fifteen; the other half will have symptoms of eczema throughout their adult lives. It is unusual for a person to develop eczema for the first time after age thirty unless they are working in a harsh climate or a wet environment.

Eczema is a very common skin disorder, affecting people in all racial and ethnic groups. The National Institutes of Health (NIH) estimates that about 15 million people in the United States have some form of eczema. The rate in adults is about 0.9 percent. It is slightly more common in women than in men.

Eczema almost never causes death; however, it is a severe psychological and economic burden to patients and their families. Flare-ups may cause children to miss school and eventually fall behind their classmates. Among adults, eczema frequently leads to taking time off from work. The National Institutes of Health estimate that U.S. health insurance companies spend more than \$1 billion per year on eczema; this is a figure comparable to the health care costs of asthma and diabetes.

# **Causes and Symptoms**

The causes of eczema are a matter of debate among doctors. For many years it was thought that eczema is primarily an allergic disease that leads to skin dryness and rashes. Doctors maintained that eczema develops from an immune overreaction inside the body that leads to inflammation and cracked, itchy skin. The breaks in the skin then let in more allergens, irritants, and microbes that made the skin itch and burn even more. The theory was supported by the observation that patients with eczema often develop other atopic ailments such as asthma, food allergies, and hay fever.

In 2006, however, a researcher in Scotland found that children with chronic eczema have a defective gene for filaggrin, a protein in the skin that normally holds in moisture. The researcher discovered that between a third and a half of his patients had a defective filaggrin gene on chromosome 1. A genetic factor may help to explain why eczema has increased in developed countries since the 1980s; air pollution and the widespread use of air conditioning and central heating could further dry out skin that is already dry and fragile, allowing irritants to enter and trigger the inflammation that characterizes eczema. On the other hand, many patients with eczema do not have the defective filaggrin gene, while others with the gene do not develop eczema.

At one time it was thought that emotional stress caused eczema. It is now known that while stress can make a flare-up of eczema worse, it does not cause the disorder.

The symptoms of eczema may include:

- Areas of dry, leathery, or discolored skin.
- Intense itching in the affected areas.
- Blisters that ooze tissue fluid and then crust over.
- Rash. In children, the rash is most often found on the face, elbows, and knees; in adults, it is more common on the skin inside the knees and elbows.
- Raw areas of skin from scratching.
- Reddened or inflamed skin around the blisters.
- Bacterial infections that develop in the broken skin.

# Diagnosis

The doctor will usually base the diagnosis on the basis of an examination of the patient's skin, together with a personal and family history of atopic disorders. The patient may be referred to a dermatologist (a doctor who specializes in disorders of the skin) to rule out the possibility that the skin problems are caused by other diseases. The dermatologist may take a skin biopsy in order to exclude other causes, but this type of test is not necessary to make the diagnosis.

# Treatment

Treatment of eczema focuses on reducing itching (and therefore scratching); lowering inflammation; and preventing flare-ups. The specific medications that are used depend somewhat on the severity of the patient's symptoms.

Common treatments include:

- Moisturizers. Lubricating creams or ointments should be applied to the skin after bathing or showering to seal moisture in the upper layer of skin cells. The most effective moisturizers are those that contain petrolatum (petroleum jelly).
- Creams or powders that contain cortisone. Different formulations may be tried, as some people are allergic to the preservatives used to make the creams. These products should be discontinued during remissions because long-term use can cause skin irritation or discoloration, thinning of the skin, infections, and stretch marks on the skin.
- Severe eczema may be treated with oral antihistamines or oral steroid medications. The steroid drugs are effective in relieving itching but, like cortisone creams, they should not be used for long periods of time because of potentially serious side effects. These side effects include cataracts, osteoporosis, muscle weakness, lowered resistance to infections, high blood pressure, and thinning of the skin.
- Antibiotic creams or oral antibiotics. These may be prescribed if the patient develops a bacterial infection in broken or inflamed skin.
- Immunomodulators. This newer, prescription-only class of drugs is sometimes recommended for children over two years of age and adults. The Food and Drug Administration (FDA) issued a warning in 2006, however, that these drugs should be used only in patients who are not helped by other treatments, because of the risk of long-term damage to the immune system.
- Phototherapy. Phototherapy is the use of natural sunlight or ultraviolet light. While some patients benefit from phototherapy, it also speeds up aging of the skin and increases the risk of skin cancer. Patients who want to try this approach to treating eczema should consult their doctor first.

Alternative treatments that are sometimes recommended for eczema include evening primrose oil, vitamin therapy, various Chinese herbal

# WORDS TO KNOW

- **Atopy:** The medical term for an allergic hypersensitivity that affects parts of the body that are not in direct contact with an allergen. Hay fever, eczema, and asthma are all atopic diseases.
- **Dermatitis:** The medical term for inflammation of the skin.
- **Filaggrin:** A protein in the skin that is defective or lacking in some patients with eczema.
- **Phototherapy:** The use of sunlight or ultraviolet light to treat eczema or other skin disorders.
- **Remission:** A period in the course of a disease when symptoms disappear for a time.

medications, and nutritional supplements, though none of these have been scientifically proven effective.

# Prognosis

About half of children with eczema will have relatively clear skin and few flare-ups as adults. The other half are likely to be troubled with recurrent eczema in adult life.

# Prevention

There is no cure for eczema. Treatment largely consists of identifying the specific substances that trigger skin reactions, avoiding contact with them whenever possible, and applying products that relieve the itching, dryness, and inflammation of the skin.

# The Future

Eczema is becoming a more common disease in the developed parts of the world, with rates as high as 18 percent in children of some countries. It is also rising among people who migrate from less developed to more developed countries. It may be possible in the future to develop medications that would stimulate filaggrin production in patients with a defective gene for this skin protein. Another area of research is focused on the benefits of early protective skin care in children with eczema. Some doctors think that early treatment of the skin condition may prevent hay fever, food allergies, and asthma from developing in later childhood, or at least reduce their severity.

SEE ALSO Asthma; Hay fever

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# Edwards Syndrome

# Definition

Edwards syndrome is a genetic disorder caused by the presence of an extra copy of chromosome 18 or by a portion of chromosome 18 translocated (attached) to another chromosome plus two copies of chromosome 18. It is the second most common genetic disorder caused by a trisomy, the most common being Down syndrome, or trisomy 21. Edwards syndrome was first identified in 1960 by a British geneticist,

Also Known As Trisomy 18

### Cause

A third copy of chromosome 18

### Symptoms

Mental retardation, failure to thrive, heart defects, seizures, early death

### Duration

Lifelong



John H. Edwards (1928–2007), who analyzed blood samples taken from a severely malformed baby who died in a British children's hospital.

# Description

Edwards syndrome is a condition in which babies who survive until birth have a low birth weight; small and abnormally shaped heads; small jaws and mouth; and clenched fists with overlapping fingers. They also suffer from mental retardation, heart defects, kidney disorders, and other organ abnormalities affecting most systems of the body. Most children with Edwards syndrome die within a few days of birth.

# **Demographics**

Edwards syndrome occurs once in every 6,000 to 8,000 live births in the United States. About 95 percent of children conceived with the syndrome, however, die before birth. Between 5 and 10 percent of children born with trisomy 18 live beyond twelve months.

About 80 percent of children born with Edwards syndrome are female; it is thought that being male increases the risk of dying from

Image showing three copies of chromosome 18 (in blue), resulting in Edwards syndrome. © DEPARTMENT OF CLINICAL CYTOGENETICS, ADDENBROOKES HOSPITAL/SCIENCE PHOTO LIBRARY/PHOTO RESEARCHERS, INC.

# **Perinatal Hospice**

A heartbreaking aspect of prenatal testing is that some parents are told that their child has Edwards syndrome or another condition that will lead to the baby's death shortly after birth. Parents who choose not to end the pregnancy when they receive this diagnosis can now turn to hospice programs that will support them and help them make plans for the birth and other medical decisions. Although hospices for dying adults and school-age children have existed in the United States since the 1970s, until the early 1990s there were few hospice programs available for babies with fatal conditions.

Since the 1990s at least 40 U.S. hospitals have started perinatal hospice programs, which help families make decisions about the birth, to take birthing classes apart from families expecting healthy babies, to prepare religious and other rituals for the child's birth, and to stay with the baby during its short life. If the child lives beyond the first few days, the parents are given advice on caring for the child at home. Families who have used perinatal hospice programs are very positive about the programs, according to one group of researchers. Parents report being more emotionally and spiritually prepared for their infant's death; experiencing less intense despair and sadness; and feeling a sense of peace.

the disorder before birth. The risk of having a child with the disorder increases with the mother's age.

Edwards syndrome is equally common in all races and ethnic groups.

# **Causes and Symptoms**

Almost all cases-92 percent-of Edwards syndrome involve genetic errors that occur during the formation of germ cells (eggs and sperm) or during cell division shortly after the egg is fertilized by the sperm. The most common form of Edwards syndrome occurs when an egg or sperm carrying two copies of chromosome 18 is involved in conception. The reason for the extra copy in the abnormal germ cell is a genetic error called nondisjunction. During the normal process of germ cell formation, the paired chromosomes in the cell divide so that each daughter cell has only one member of the pair. In nondisjunction, one daughter cell gets both members of the chromosome pair and the other cell has none. If a germ cell carrying two copies of chromosome 18 is fertilized by a normal germ cell from the other parent, the child will have three copies of chromosome 18. This genetic error is called a full trisomy 18.

A few cases (about 2 percent) of Edwards syndrome develop when a part of chromosome 18 becomes attached to another chromosome

either before or at the moment of conception. This type of genetic error is called a translocation. The child will be born with two copies of chromosome 18 plus some extra genetic material from chromosome 18 attached to another chromosome. A child with this type of genetic error is said to have partial trisomy 18.

About 5 percent of cases of Edwards syndrome occur in children who have some body cells with the extra copy of chromosome 18 and some body cells without the extra copy. This condition is called mosaic trisomy 18. It is thought to result from random errors in cell division during the early stages of fetal development. Children with mosaic trisomy 18 are less severely affected than those with full trisomy 18 and have a longer survival period.

The symptoms of Edwards syndrome include a high mortality rate even before birth. Most embryos with full trisomy 18 die during pregnancy or are expelled from the mother's womb in what is called a spontaneous abortion or miscarriage. Those who survive until birth usually live only a few days; most die from breathing problems, heart or kidney defects, generalized infection, or feeding difficulties. Although some children with mosaic trisomy 18 do not have all the physical features that characterize the syndrome, the following are considered typical:

- Mental retardation and delayed movement (100 percent of infants)
- · Poor muscle tone, seizures, and breathing difficulties
- Abnormally small head, abnormally small eyes, flattened nose, small jaw, pointed ears, cleft lip, and cleft palate
- Severely retarded growth with clenched hands, missing bone in forearm, rocker-bottom feet
- Heart abnormalities (90 percent of infants), underdeveloped lungs, incomplete esophagus, double spleens, missing appendix, and missing gallbladder
- Underdeveloped sex organs in both boys and girls
- Underdeveloped thyroid and thymus glands

# Diagnosis

Diagnosis of Edwards syndrome is usually made on the basis of the child's appearance at birth. It can be diagnosed before birth on the basis of ultrasound studies during the first three months of pregnancy; a sample of the mother's blood plasma; or by genetic analysis of cells taken from the baby's blood or the amniotic fluid that surrounds the baby inside the womb.

In some cases the diagnosis is suspected before birth on the basis of certain characteristics of the pregnancy. These include a smaller than average placenta (the organ that attaches the baby to the wall of the mother's uterus), a low level of fetal activity, irregular heartbeat, and slowed fetal growth.

### Treatment

There is no treatment for Edwards syndrome itself. Infants who survive the first two days often require intensive treatment for infections, breathing difficulties, and problems with feeding. Surgery to correct heart defects is rarely performed because of the baby's short life expectancy.

# Prognosis

The prognosis of Edwards syndrome is extremely poor. Newborns have a 40 percent chance of living a full month; infants have a 5 percent chance of surviving a full year; children have a 1 percent chance of surviving to age ten years. The very small number of children with Edwards syndrome who live as long as ten years suffers from severe mental retardation, digestive problems, and difficulty in walking.

## Prevention

Since Edwards syndrome is thought to be caused by a spontaneous genetic mutation rather than an inherited genetic defect, there is no way to prevent it. Pregnant women over thirty-five should have tests during the first trimester (three-month period) of pregnancy to screen for the syndrome. These tests may involve ultrasound studies, which can detect abnormalities in the baby's heart or facial development, followed by a photographic analysis of cells taken from the fluid that surrounds the baby in the womb. This analysis, or karyotype, is needed to distinguish Edwards syndrome from other genetic disorders that can cause heart defects and facial abnormalities. Doctors recommend that the parents of a child with Edwards syndrome should consult a genetic counselor for advice about future pregnancies.

# The Future

It is possible that the increasing numbers of women having children in their thirties or forties will lead to an increase in the number of children born with Edwards syndrome. Many women whose fetuses are diagnosed with the syndrome choose to end their pregnancies before childbirth. A newer option is the perinatal hospice (see sidebar), a special type of hospital program where parents who know that their baby has a fatal condition can choose to have the baby and remain with it during the baby's few days of life.

# WORDS TO KNOW

- **Germ cell:** A cell involved in reproduction. In humans the germ cells are the sperm (male) and egg (female). Unlike other cells in the body, germ cells contain only half the standard number of chromosomes.
- **Hospice:** A facility or program for meeting the spiritual as well as the physical needs of people who are terminally ill.
- **Karyotype:** A photomicrograph of the chromosomes in a single human cell. Making a karyotype is one way to test for genetic disorders.
- **Mosaicism:** A condition in which a person has some body cells containing an abnormal number of chromosomes and other cells containing the normal number. Mosaicism results from random errors during the process of cell division that follows conception.
- **Nondisjunction:** A genetic error in which one or more pairs of chromosomes fail to separate

during the formation of germ cells, with the result that both chromosomes are carried to one daughter cell and none to the other. If an egg or sperm with a paired set of chromosomes is involved in the conception of a child, the child will have three chromosomes in its genetic makeup, two from one parent and one from the other.

- **Perinatal:** Related to the period around the time of a baby's birth.
- **Rocker-bottom feet:** Abnormally long and slender feet with pointed heels turned outward like the bottom rails of a rocker.
- **Translocation:** A genetic error in which a part of one chromosome becomes attached to another chromosome during cell division.
- **Trisomy:** A type of genetic disorder in which a cell contains three copies of a particular chromosome instead of the normal two.

In addition to perinatal hospice programs, there are also online support groups for parents of children with Edwards syndrome and other rare genetic disorders.

### SEE ALSO Down syndrome; Patau syndrome

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# Emerging Diseases

Emerging infectious diseases, or EIDs, are a category of contagious illnesses that have become public health concerns during the past 20 years and are expected to become more common or spread to more countries in the near future.

EIDs include several different types of infections:

- Diseases involving newly identified organisms (AIDS, SARS).
- Diseases involving mutations or changes in a known disease organism (avian flu).
- Diseases that spread to a geographical area where they were previously unknown (Lyme disease, West Nile virus, hantavirus infection).
- Diseases that have become more difficult to treat because of resistance to commonly used antibiotics (drug-resistant tuberculosis).

There are several reasons why a disease can become an EID:

- Mass travel and tourism. Diseases like SARS and AIDS spread rapidly because of the popularity and speed of air travel.
- Bioterrorism, or the intentional spread of deadly diseases as a method of warfare.
- Evolution of the disease organisms themselves.
- Breakdowns in public health, such as malnutrition, poor sanitation, or refugee situations.
- Overuse of antibiotics in treating bacterial infections.
- Importing animals that may be infected with previously unknown organisms for study in laboratories or as pets.
- Changes in human vulnerability to disease. For example, smallpox could reemerge as a disease because routine vaccination against it was discontinued in the 1970s.

**SEE ALSO** AIDS; Avian influenza; Ebola and Marburg hemorrhagic fevers; Hantavirus infection; Lyme disease; Severe acute respiratory syndrome; Tuberculosis; West Nile virus infection

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# Emphysema

# Definition

Emphysema is a form of progressive lung disease that is characterized by chronic shortness of breath and long-term disability. It is sometimes grouped together with chronic bronchitis under the name of chronic obstructive pulmonary disease, or COPD. Many people who are diagnosed with emphysema also have chronic bronchitis.

# Description

Emphysema is a lung disease in which a person's ability to breathe easily and deeply is steadily weakened over time by the destruction of lung tissue. The human lung consists of tissue containing millions of tiny air sacs called alveoli, which are arranged like bunches of grapes around very

#### Also Known As

Chronic obstructive pulmonary disease, COPD

### Cause

Damage to air sacs in lungs due to smoking, air pollution, hereditary factors

#### Symptoms

Shortness of breath, coughing, wheezing, inability to take a deep breath

#### Duration

Lifelong after onset

Image of a patient's lung with emphysema. The tiny air sacs become enlarged and may collapse. DR. GARY GAUGLER / PHOTO RESEARCHERS, INC.



small air tubes called bronchioles. There are about 300 million alveoli in each lung. When a person breathes in air, the air travels from the nose and mouth through the windpipe and then into the right and left bronchi, which are the main air passages into each of the two lungs. The bronchi divide and subdivide repeatedly into smaller and smaller air passages, finally ending in the bronchioles and alveoli.

In a healthy person, oxygen from the air that has been taken in is exchanged in the walls of the alveoli for carbon dioxide in the person's blood. When the person breathes out, the carbon dioxide leaves the body in the air that travels out from the lungs and through the upper airway to the nose and mouth.

To perform its function effectively, the tissue in the lungs that separates the alveoli from one another needs to be as elastic as possible. The alveoli contain tiny elastic fibers in their cell walls that allow them to act like miniature balloons. What happens in emphysema is that smoke or other irritants causes the alveoli to become inflamed and lose their elasticity. The bronchioles start to collapse, which traps air inside the alveoli and overstretches them. In time the alveoli rupture, leading to the formation of fewer but larger air sacs in the lungs. These larger and less flexible sacs are less efficient in forcing air out of the lungs when the person breathes out. As a result, the person has to breathe more frequently or

# The High Cost of Emphysema

Emphysema has claimed the lives of many noteworthy people in the twentieth and twenty-first centuries:

- Actors: Tallulah Bankhead, Paulette Goddard, Robert Mitchum, Vincent Price, Barbara Stanwyck, Spencer Tracy
- Entertainers: Johnny Carson, Arthur Godfrey, Dean Martin, Allan Sherman
- Musicians: Leonard Bernstein
- Politicians and public figures: William O'Neill, Patricia Nixon (wife of President Richard Nixon), Coleman Young
- Writers and poets: Samuel Beckett, William F. Buckley, Jr., Erskine Caldwell, T. S. Eliot, Robert Heinlein



Actress Barbara Stanwyck. SILVER SCREEN COLLECTION/ HULTON ARCHIVE/GETTY IMAGES.

breathe harder in order to get enough oxygen and get rid of carbon dioxide.

In addition to the loss of elasticity in the alveoli, the cells in the airways secrete more mucus than usual, which collects in the airways and clogs them, making breathing even more difficult.

# Demographics

Emphysema is increasing in the United States and other developed countries primarily because of cigarette smoking. It is almost entirely a disease of adults. About 12 million adults in the United States have been diagnosed with the disease; however, many doctors believe emphysema is underdiagnosed. Between 4 and 6 percent of male adults and 1 to 3 percent of female adults are estimated to have emphysema. The number of women diagnosed with the disease is rising rapidly; the year 2000 was the first year that more women than men were identified as having emphysema. About 120,000 people die each year in the United States from emphysema. People who develop emphysema as a result of smoking generally start to have symptoms in their late forties or early fifties. Those who have emphysema because of a genetic condition (described later) may begin to have symptoms in their thirties. This genetic condition, known as alpha 1-antitrypsin deficiency, is more common in Caucasians than in members of other races and accounts for about 2 percent of all emphysema cases in the United States.

Some people who do not smoke cigarettes are at increased risk of emphysema. They include:

- People who work in occupations that expose them to high levels of dust from grain or cotton, or chemical irritants. These occupations include mining, certain types of agricultural work, and lumbering.
- People exposed to high levels of automobile exhaust or secondhand smoke.
- People with certain diseases that affect connective tissue, such as Marfan syndrome.

### **Causes and Symptoms**

Emphysema is caused by a weakening of the tissues in the lungs as a result of inflammation due to smoke or other chemical irritants in the air, or a hereditary deficiency of a protein that protects the elasticity of lung tissue. As the bronchioles in the lungs collapse and the alveoli become enlarged, the lungs become less efficient in getting rid of carbon dioxide and the person has to breathe more frequently in order to get enough oxygen. In addition, the person has to use his or her chest muscles to expel air from the lungs forcefully rather than being able to rely on the normal movement of the diaphragm during breathing. This need to use muscular force leads to the development of a so-called barrel chest; that is, the person's chest is almost the same size from front to back as from side to side.

About 2 percent of cases of emphysema are caused by a deficiency of a liver enzyme known as alpha 1-antitrypsin, or A1AT. The enzyme ordinarily protects the alveoli in the lungs from damage by another enzyme that harms connective tissue. In people with A1AT deficiency, there is not enough of the protective enzyme to keep the alveoli in good working condition. A1AT is an inherited condition caused by a mutation in a gene on chromosome 14. In addition to shortness of breath, coughing, and wheezing, people with emphysema often develop the following symptoms:

- Pursed-lips breathing. This is a way of partially closing the lips that allows the person to fully exhale. When the mouth opening is smaller, the airways that have been weakened by the disease open wide and allow the person to expel more air from the lungs.
- Greater difficulty exercising or doing work that requires physical activity.
- Loss of appetite and weight loss. Eating can make it harder to breathe because the stomach expands during a meal and pushes upward against the diaphragm.
- Fatigue. Emphysema leads to a lower level of oxygen in the blood, which in turn causes people to feel tired easily.
- Slow recovery from such upper respiratory infections as colds and flu.

It is important to note that emphysema is a disease that develops gradually; thus its symptoms may take years to become bothersome enough to send the patient to their doctor.

# Diagnosis

Emphysema is most often diagnosed by pulmonary function tests or PFTs. These are tests in which the patient is asked to breathe into a device called a spirometer. The spirometer measures how much air the patient's lungs can hold and how fast the air moves in and out of the lungs. It can also be used to determine how well the lungs are exchanging oxygen and carbon dioxide. Another way the spirometer can be used is to test the effectiveness of inhaled medications in treating the patient's emphysema.

Patients are also typically given a chest x ray or computed tomography (CT) scan to look for damage to lung tissue and the possible presence of a lung infection. To measure the amount of oxygen and carbon dioxide in the patient's blood, he or she may be given a type of blood test called the arterial blood gases test. This test can help the doctor evaluate the severity of the patient's emphysema.

# Treatment

There is no cure for emphysema. Treatment is focused on slowing the progress of the disease and easing the patient's symptoms. The first part of treatment for patients who smoke is to quit the habit.

The next stage in treatment is the use of medications. The doctor may prescribe one or more of the following types of drugs:

- Bronchodilators. Bronchodilators are drugs that work by opening up the airways, which allows for more efficient exchange of carbon dioxide and oxygen. Some are taken in tablet form while others are dispensed in inhalers. Depending on the severity of the person's emphysema, patients may use the inhaler only when needed for shortness of breath or they may take a dose of the medication at prescribed regular intervals.
- Steroids. This type of medication works by lowering the inflammation in the tissues lining the airways. These drugs can also be taken in pill form or through inhalers.
- Antibiotics. People who have infections in the lungs as well as emphysema may be given antibiotics to treat the infections.
- Oxygen. Patients who have severe attacks of emphysema may need to go to the hospital for oxygen therapy. There are also oxygen tanks that can be used in the home; some of these are portable units.

Emphysema is sometimes treated surgically. In some cases, part of the diseased lung is removed. This procedure creates space for the remaining portions of the lungs; it does improve breathing and quality of life for some patients. Another surgical option is lung transplantation. This is a risky procedure, however, and requires the patient to take medications to prevent the rejection of the transplanted lung. In addition, not everyone qualifies for transplantation, and those who do are limited by the short supply of available organs.

Another important part of treatment for emphysema is called pulmonary rehabilitation. It is aimed at educating patients about their disease and helping them with lifestyle changes that will slow the progression of the disease and improve quality of life. Pulmonary rehabilitation includes a physical exercise program designed to improve the patient's physical endurance and energy level. Many patients are also encouraged to lose weight in order to reduce the burden on their lungs. One important benefit of pulmonary rehabilitation is psychological: Patients report that their self-esteem and sense of control over their life improve when they start to see benefits from the rehabilitation program.

# WORDS TO KNOW

- Alveolus (plural, alveoli): The medical term for one of the tiny air sacs in the lungs where oxygen is transferred from the lungs to the blood and carbon dioxide is removed.
- **Bronchiole:** A very small thin-walled air passage in the lungs that branches off from a bronchus.
- **Bronchus (plural, bronchi):** One of the two major divisions of the airway that lead into the right and left lungs.
- **Diaphragm:** A sheet of muscle tissue that divides the chest cavity from the abdominal cavity.
- **Progressive:** A term that refers to a disease that gets worse over time.
- **Spirometer:** A device that is used to test the air capacity of a person's lungs and the amount of air that enters and leaves the lungs during breathing.

# Prognosis

Emphysema is known to shorten a patient's lifespan. It is the fourth most common cause of death in the United States as of 2008, being responsible for 4.5 percent of all deaths and a contributing factor in another 4.3 percent. Some patients, however, live longer than others depending on the cause of their emphysema and the measurement of their lung capacity at diagnosis. Patients who have smoked twenty cigarettes per day for twenty years or longer with a severely reduced breathing capacity have the worst prognosis; only 5 percent survive for twelve years after diagnosis.

### Prevention

Most cases of emphysema can be prevented by simply not smoking or by quitting smoking as soon as possible and avoiding secondhand smoke. Emphysema related to genetic factors cannot always be prevented, but its development can be postponed in people who inherited the defective gene by avoiding smoking.

### The Future

Emphysema is likely to continue to rise in the United States because of the numbers of long-term female smokers who are now getting to the age when the symptoms of emphysema appear. In addition, the rates of emphysema around the world are predicted to rise rapidly over the next few decades as more and more people take up smoking. It is possible that more effective drugs will be developed to treat emphysema, but a cure for the disease is unlikely in the near future.

SEE ALSO Bronchitis; Marfan syndrome; Smoking

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# Encephalitis

# Definition

Encephalitis is a severe infection or inflammation of the brain itself. It should be distinguished from meningitis, which is an inflammation of the protective membranes that cover the brain and line the spinal cord. However, the two conditions can exist together. Encephalitis is usually caused by viruses, but can also result from infections caused by bacteria, fungi, or parasites.

# Description

Encephalitis is an illness that can range from mild cases with flu-like symptoms to life-threatening, severe cases that can kill patients or leave them with permanent problems. The severity of the illness varies according to the patient's age, the organism that is causing the encephalitis, and the condition of the person's immune system. Mild viral encephalitis may have no symptoms at all, or a brief period of headaches, tiredness, and irritability.

Severe encephalitis often begins with a prodrome, or a period of warning symptoms. After the prodrome, victims of the disease may become feverish, vomit, become drowsy, find that their eyes are sensitive to light, complain of a stiff neck, or feel unsteady on their feet when they try to walk. They should see their doctor at once. If they lose their memory, have speech problems, are paralyzed, or go into a coma, they need emergency medical treatment and should be taken to the hospital as soon as possible.

# **Demographics**

The exact number of cases of encephalitis in the United States in an average year is difficult to estimate because mild cases may not be diagnosed, and the reporting of severe cases is not standardized. Between 1,500 and 3,000 cases of viral encephalitis are reported to the Centers for Disease Control and Prevention (CDC) each year, but most doctors think that the actual number of cases is higher. According to the CDC, it costs about \$150 million each year to control the mosquitoes that spread some types of viral encephalitis.

Also Known As Inflammation of the brain

### Cause

Viruses (in most cases); also bacteria, fungi, and parasites

### Symptoms

Mild cases: vague flu-like symptoms; severe cases: sudden fever, headaches, seizures, vomiting, confusion

Duration

Two to three weeks



Some statistics for specific types of viral encephalitis are as follows:

- Japanese encephalitis: 30,000–50,000 cases each year in Asia; about one case per year in Americans traveling in Asia.
- Eastern equine encephalitis, found in the eastern United States: five cases per year on average.
- La Crosse encephalitis, found in California and the Great Lakes region: seventy cases per year on average.
- West Nile virus: about 1,200 cases per year in the United States.
- Herpes simplex encephalitis, caused by the virus that causes cold sores: about one person in every 250,000 per year in the United States.
- Encephalitis following measles: about one case per 1,000 infected persons.
- Encephalitis following chickenpox: one case in every 2,000 infected persons.
- Encephalitis following rabies: one to three cases in the United States each year.

# **Causes and Symptoms**

Most cases of encephalitis in the United States are caused by viruses:

- Herpes simplex virus (HSV) 1 and 2. This is the virus responsible for cold sores and genital herpes.
- Poliovirus.
- Arboviruses. The arboviruses are a family of viruses transmitted to humans by infected mosquitoes, ticks, or other blood-sucking insects. These types of encephalitis include West Nile virus, Japanese encephalitis, equine encephalitis, La Crosse encephalitis, and St. Louis encephalitis.
- The rabies virus.

Toxoplasmosis, a disease caused by a parasite, and Lyme disease, a bacterial infection, can cause encephalitis.

Image of a child's brain with encephalitis caused by the herpes virus. Pink and blue areas are inflammation caused by the infection. AIRELLE-JOUBERT / PHOTO RESEARCHERS, INC. Symptoms of encephalitis in infants may include:

- Vomiting
- Body stiffness
- Crying that increases when the baby is picked up or held
- A fullness or bulge in the soft spot at the top of the baby's head

In addition to the symptoms of encephalitis in adults discussed earlier, people with severe infections may also have:

- Visual hallucinations
- Seeing double
- · Personality changes
- Loss of sensation in some parts of the body
- Impaired judgment
- Muscle weakness

# When to See the Doctor

A person who has been exposed to organisms that can cause encephalitis combined with symptoms of the illness should see their doctor quickly if:

- The person has cold sores or sores around the genitals following close contact with another person.
- He or she has recently had measles or chickenpox, or been exposed to someone with those diseases.
- The person knows that he or she has recently been bitten by a tick.
- He or she has been bitten by a bat, raccoon, dog, or coyote.
- The person has been in wooded, damp, or marshy areas where mosquitoes are likely to breed.
- He or she has traveled to foreign countries where various types of encephalitis are common, particularly eastern Asia.

# Diagnosis

The patient's history of recent travels, exposure to animals or infected persons, or exposure to

the herpes simplex virus or arboviruses are important clues in diagnosis. If the patient is unconscious or cannot speak, family members may be asked for information about his or her recent activities.

The specific diagnostic tests that may be done depend partly on the type of disease organism that the doctor suspects is causing the encephalitis. Some common diagnostic tests include:

- Blood tests. Blood tests can be used to diagnose West Nile virus
- A neurological examination. This examination allows the doctor to evaluate the patient's memory and other mental functions, sight and hearing, reflexes, gait and movement, mood, level of consciousness, and other functions of the nervous system that may be affected by encephalitis.
- Spinal tap. In a spinal tap, a small sample of spinal fluid is withdrawn through a needle inserted into the lower back. It can be

tested for evidence of a bacterial or fungal infection. In some cases, a spinal tap can also be used to diagnose HSV encephalitis.

- Electroencephalogram (EEG). An EEG is a test that measures patterns of electrical activity in the brain. HSV encephalitis produces a telltale abnormal EEG pattern.
- Imaging studies of the brain. Computed tomography (CT) scans and magnetic resonance imaging (MRI) are the types of imaging studies most frequently used to diagnose encephalitis.
- Brain biopsy. In a brain biopsy, a surgeon removes a small sample of brain tissue to analyze it for the presence of HSV. This test is rarely done except when brain imaging or other tests for HSV do not provide definite results.

# Treatment

Treatment of mild encephalitis usually includes bed rest at home, drinking lots of fluids, and taking over-the-counter pain relievers to bring down fever and treat headaches. If the patient gets worse suddenly, or if the infection is severe from the start, emergency treatment in a hospital is necessary.

Specific forms of treatment for encephalitis in the hospital may include:

- Acyclovir for HSV encephalitis. Acyclovir is an antiviral drug that is given intravenously when the doctor suspects HSV encephalitis. It is given for a period of two to three weeks.
- If the encephalitis is caused by a bacterium, the doctor may prescribe antibiotics. Antibiotics cannot be used to treat encephalitis caused by viruses, however.
- Patients are monitored for signs of swelling of the brain, and they are given intravenous fluids to prevent dehydration.
- Patients who are having seizures are given anticonvulsants (antiseizure drugs).
- Sedatives may be given to patients who are restless or having trouble sleeping.
- Patients who are partially paralyzed or having trouble breathing are put on a respirator.

Patients who have had severe encephalitis may suffer permanent damage if they survive. They may need physical therapy, speech therapy, or psychological counseling after the acute illness has passed.

# Prognosis

The prognosis for recovery from encephalitis depends on the virus or other disease agent that is causing the infection, the patient's age, and his or her general state of health. In general, infants and the elderly are at the greatest risk of death from encephalitis.

People with mild cases of encephalitis usually recover without long-term complications.

The prognoses for specific types of encephalitis are as follows:

- Rabies encephalitis: 100 percent mortality.
- Japanese encephalitis: 60 percent mortality, usually within the first week of illness.
- Untreated HSV encephalitis: 50–75 percent of patients die within eighteen months. The fatality rate drops to 20 percent in patients treated with acyclovir. Forty percent of survivors have long-range learning disabilities, epilepsy, movement disorders, memory loss, and psychiatric problems.
- St. Louis encephalitis: 30 percent mortality.
- West Nile virus: 10 percent mortality.

# Prevention

Japanese encephalitis is the only encephalitis for which there is an effective vaccine for humans. People planning a trip to Japan or eastern Asia may want to consider being vaccinated before departure. There was a vaccine for horses against West Nile virus but not one for humans as of late 2008.

Other preventive measures to lower one's risk of encephalitis include:

- Draining water from bird baths, wading pools, and other outdoor water containers when not in use.
- Using insect repellent when outdoors; applying it to exposed skin and spraying clothing with repellent.
- Wearing long-sleeved shirts and long pants on hikes or other outdoor activities when the weather permits.
- Keeping window and door screens in good repair so that mosquitoes cannot get inside the house.
- Covering baby strollers or carriers with mosquito netting when taking the baby for a walk outdoors.

# WORDS TO KNOW

Arboviruses: A family of viruses spread by bloodsucking insects.

Gait: A person's manner of walking.

**Meningitis:** Inflammation of the membranes that cover the brain and line the brain and spinal cord.

**Prodrome:** A period before the acute phase of a disease when the patient has some characteristic warning symptoms.

- Having pet dogs and cats vaccinated against rabies, and avoiding bats, raccoons, and other wild animals that are acting aggressive.
- Avoiding sexual contact with people infected by genital herpes.
- Avoiding kissing or sharing drinking glasses with people with cold sores.
- Having children vaccinated against diseases such as measles and mumps.
- Seeking prompt medical treatment for any high fever associated with an infection.

### The Future

Researchers are working on developing vaccines against West Nile virus and other mosquito-borne viral infections. As of 2008, the National Institutes of Health was testing a recently developed vaccine against equine encephalitis, and a new drug related to acyclovir as a treatment for HSV encephalitis.

**SEE ALSO** Chickenpox; Cold sore; Genital herpes; Lyme disease; Measles; Meningitis; Rabies; Toxoplasmosis; West Nile virus infection

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Epilepsy See Seizure disorder.

# **Epstein-Barr Infection**

See Chronic fatigue syndrome.

# F



Also Known As FAS; alcohol in pregnancy

### Cause

Maternal alcohol intake during pregnancy

### Symptoms

Characteristic facial features, slowed growth, brain and spinal cord abnormalities

Duration

Lifelong



# Fetal Alcohol Syndrome

# Definition

Fetal alcohol syndrome (FAS) is defined as the most severe form of fetal alcohol spectrum disorder, or FASD. It is characterized by abnormalities in facial features, slowed growth, and abnormalities in the structure and functioning of the brain and spinal cord.

# Description

Fetal alcohol syndrome is a disorder that affects a child's ability to learn, to pay attention, and to control his or her behavior as well as to grow normally. It is the leading known cause of mental retardation in North America. Children with FAS are undersize and underweight for their age, and they have smaller than average heads as well as certain facial features associated with the disorder. The physical features of FAS are usually evident at birth, but problems with learning and other abnormalities in brain development may not be detected until the child starts school. The child's learning and behavioral problems persist over his or her lifetime, while the child's facial features usually become less noticeable as he or she grows into adolescence and adulthood.

# Demographics

Researchers estimate that about 1 percent of the general U.S. population is affected by fetal alcohol spectrum disorders and that 40,000 babies are

#### Fetal Alcohol Syndrome



Nurse working with a boy who has fetal alcohol syndrome. These patients often have small eye openings and other facial abnormalities. AP IMAGES.

born every year with some kind of birth defect caused by the mother's drinking. However, fetal alcohol syndrome itself is the only disorder in this group that has been formally defined by the Centers for Disease Control and Prevention (CDC) and by the International Classification of Diseases (ICD). Fetal alcohol syndrome is estimated to occur in one or two out of every 1,000 live births in the United States. The rate may be higher in some countries where binge drinking is common. The cost of treating one child with FAS is estimated to be \$2 million over his or her lifetime. The cost of FAS to the American public runs to more than \$321 million each year.

As far as is known, FAS affects boys and girls equally and affects children of all races equally. The primary risk factor is the amount and frequency of the mother's drinking. Other risk factors in mothers include being over age thirty, a history of heavy drinking, and low social or economic status. Smoking and the use of other drugs can worsen the effects of alcohol on the fetus.

# **Causes and Symptoms**

Fetal alcohol syndrome occurs as the result of a woman drinking alcohol during her pregnancy. Alcohol is a teratogen, which means that it is a substance that causes birth defects. Although heavy or binge drinking increases the risk of having a child with FAS, even moderate or occasional drinking can affect the unborn baby. In addition, it does not matter what type of alcoholic beverage is consumed. Beer or wine can harm the baby as much as whiskey or other forms of distilled alcohol. It also does not matter whether the mother drinks throughout her pregnancy or only during a brief period. Alcohol can affect the baby's development at any point during pregnancy.

The ways in which alcohol affects the development of the baby before birth are not fully understood. What is known is that the fetus depends on the mother's liver to clear alcohol from its bloodstream as well as her own. It is thought that alcohol and a chemical called acetaldehyde, which is formed during the body's digestion of alcohol, disrupt the process of cell reproduction and tissue formation in the baby's body. In addition, these chemicals interfere with the transfer of vitamins, minerals, and other nutrients from the mother's body to the baby, thus starving the baby of nutrients that are vital to healthy growth. Last, alcohol and acetaldehyde interfere with the oxygen supply to the baby.

Some pregnancies end in miscarriage if the mother is a heavy drinker. A child with FAS may also be born prematurely or die of sudden infant death syndrome (SIDS).

The symptoms of FAS at birth may include withdrawal syndrome if the mother had been

# **Did You Know?**

Although fetal alcohol syndrome was not named until the twentieth century, people in ancient times noticed a connection between a pregnant woman's drinking and health problems in her baby. Ancient Greek and Roman doctors as well as passages in the Bible referred to cases of what would now be identified as FAS. In 1899 a British doctor who treated pregnant women in prison noticed that those who drank were more likely to lose their babies before delivery than those who did not drink alcohol. William Sullivan was the first doctor in modern times to suggest a connection between alcohol use and bad outcomes in childbirth.

In 1968 French doctor Paul Lemoine published an article about the unusual facial features of children born to alcoholic mothers. Other researchers in the United States and Sweden confirmed Dr. Lemoine's findings. In 1973 FAS was named by two American researchers, Kenneth Jones and David Smith, at the University of Washington Medical School in Seattle. Although some doctors were initially skeptical that alcohol could cause so much damage to unborn children, researchers used studies on monkeys and apes to prove that alcohol is indeed harmful to a child's development before birth.

drinking heavily shortly before delivery. Newborns suffering from alcohol withdrawal may be hyperactive, restless, or suffer from seizures. The physical and developmental symptoms are described in the next section because they are used to diagnose the disorder.

# Diagnosis

Diagnosis of fetal alcohol syndrome is usually based on a combination of the mother's drinking history (if known) and a physical examination of the child. The Institute of Medicine (IOM) has established the following guidelines for diagnosing fetal alcohol syndrome:

- Facial features that include a smooth upper lip (lacking the normal indentation between the nose and the mouth); a thin red border along the upper lip; and an abnormally short distance between the inner and outer corners of the eye. Other facial features may include flattening of the middle part of the face, drooping of the eyelids, and unusually small eyes. Although these facial features do not look like major deformities to most people, they tell the doctor that the child's brain has not developed normally.
- Evidence of slow growth either before or after birth. The child is below the tenth percentile in weight or height for his or her age.
- Evidence of abnormalities in the central nervous system. These may include an abnormally small head, developmental delays, learning difficulties, seizures or epilepsy, mental retardation, hyperactivity, irritability in infancy, and poor judgment or impulse control in later childhood. Many children with fetal alcohol syndrome do not learn to communicate normally with others. They may also have problems with memory and with paying attention. Some have difficulties with writing or other skills involving hand-to-eye coordination.
- Confirmed or suspected drinking by the mother.

Children with FAS may have one or more of the following physical features or characteristics even though these are not used to diagnose the syndrome:

- Cleft palate
- Heart defects
- Nearsightedness or other eye disorders
- Hearing loss
- Abnormal curvature of the spine
- Unusual creases on the palms of the hands that resemble hockey sticks
- Joints that are easily dislocated

# Treatment

Treatment of children with FAS includes treatment for birth defects at the time of birth and educational or medical treatments for learning problems or behavioral disorders as the child grows older. Behavioral disorders may be treated with medications, psychotherapy, or a combination of both.

Adults who care for or teach children with FAS should provide structure and be consistent in their behavior toward them. These children do best with routines that vary as little as possible so that they learn to see the world as a predictable place. Instructions should be brief and repeated frequently because such children have difficulty paying attention.

# Prognosis

The prognosis of a child diagnosed with fetal alcohol syndrome depends on the type and severity of the child's symptoms as well as the age at which treatment begins. Some doctors think that early diagnosis is helpful because it gets the child into treatment more rapidly.

One study followed a group of people with FAS who were between the ages of twelve and fifty-one. Researchers reported the following longterm consequences of the disorder:

- 95 percent had mental health problems.
- 55 percent had spent time in prison, a drug or alcohol treatment center, or a psychiatric hospital.
- 60 percent had been in trouble with the law.
- 82 percent were unable to live independently.
- 70 percent had problems holding a job.
- 60 percent had problems with drugs or alcohol.

# Prevention

FAS can be prevented if a pregnant woman avoids alcoholic beverages. The Institute of Medicine (IOM) recommends a prevention program on three levels: public education aimed at all women of childbearing age; selected screening programs aimed at women who drink heavily and may become pregnant; and prevention and intervention aimed at women who are heavy drinkers and have already had a child with FAS. This third level of prevention would include treatment for alcohol dependence combined with case management.

# The Future

It is likely that the number of children affected by fetal alcohol syndrome will increase in the United States over the next few decades. Reasons for
## WORDS TO KNOW

**Acetaldehyde:** A colorless liquid chemical that is produced when the body begins to digest alcohol. A chemical that causes hangovers after heavy drinking, it also contributes to fetal alcohol syndrome.

**Binge drinking:** A period of heavy drinking that lasts for two days or longer.

- **Case management:** An approach to healthcare based on personalized services to patients.
- **Syndrome:** A group of signs or symptoms that occur together and characterize or define a particular disease or disorder.
- **Teratogen:** Any substance that causes birth defects in children. Alcohol is a teratogen.

this include the easy availability of alcohol, the increasing breakdown of families and the growing number of single mothers, and the difficulty of putting preventive measures in place.

**SEE ALSO** Alcoholism; Developmental disability; Prematurity; Sudden infant death syndrome

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# **U** Fibromyalgia

## Definition

Fibromyalgia is a chronic pain disorder characterized by muscle pain throughout the person's body, general tiredness, and soreness at specific locations of the body known as pressure points. The Centers for Disease Control and Prevention (CDC) categorizes fibromyalgia as a type of arthritis; however, fibromyalgia does not cause damage to the joints or other tissues the way that other types of arthritis do.

Fibromyalgia was first defined as a disease in 1976 and accepted as a cause of workers' disability in 1987. Because fibromyalgia does not have symptoms that are unique to it, however, some doctors question whether it is really a distinctive disorder. The controversy became heated in January 2008, when Dr. Frederick Wolfe, one of the doctors who had been responsible for the diagnostic criteria drawn up by the American College of Rheumatology (ACR) in 1990, stated that he had changed his mind about the disorder. He now regards it as a physical reaction to stress, worry, and depression. Dr. Wolfe was quoted as saying, "Some of us in those days thought that we had actually identified a disease, which [fibromyalgia] clearly is not. To make people ill, to give them an illness, was the wrong thing."

## Description

Fibromyalgia is a chronic disorder characterized by widespread muscle pain, chronic fatigue, and specific areas or points of soreness on the body. It is considered a syndrome because it is a collection of signs and symptoms that occur together but cannot be traced to an identifiable cause. Most people who develop fibromyalgia are middle-aged adults. It may appear either gradually or suddenly, and may be accompanied by headaches, insomnia, and other symptoms. Most people with fibromyalgia are able to continue working, but some end up working only part-time or changing to a less demanding job.

## Demographics

Fibromyalgia can affect anyone in any age group, but it is most common in women between the ages of twenty and fifty. According to the ACR, between 3 and 6 million people in the United States have fibromyalgia,

### **Also Known As** Fibrositis, fibromyositis, fibromyalgia syndrome

### Cause

Unknown; may be partly genetic

### Symptoms

Pain throughout body, sore spots in muscles, fatigue

### Duration

May last for years



Illustration showing painful areas associated with fibromyalgia. © PHOTOTAKE INC. / ALAMY.

or one person in every fifty in the general population. Females are nine time more likely to have fibromyalgia; some of the men who are diagnosed with it are Gulf War veterans.

## **Causes and Symptoms**

Researchers do not agree about the possible causes of fibromyalgia, partly because it often occurs together with such other disorders as irritable bowel syndrome, rheumatoid arthritis, and restless legs syndrome, and partly because many do not think of it as a separate disorder in its own right. Fibromyalgia has been attributed to a number of different psychological as well as physical causes:

- Genetic factors
- A virus that affects the patient's perception of pain
- · Childhood abuse or other traumatic incidents
- Lyme disease
- High levels of substance P in the spinal fluid (Substance P is a chemical that transmits and intensifies pain signals between the brain and the rest of the body.)
- Female sex hormones
- Emotional stress
- Sleep disorders
- A buildup of excess calcium and phosphate in the muscles
- Psychological depression
- Toxic substances in the environment

In addition to muscle pain and fatigue, people with fibromyalgia may have the following symptoms:

- Headaches
- Irritable bowel syndrome
- Disturbed sleep
- Problems with memory and concentration (This symptom is sometimes called "fibro fog.")

- Chronic pain in the chest, abdomen, or pelvic region
- Changes in body weight
- Allergies; hypersensitivity to light, noises, and odors
- Shortness of breath
- Having to urinate more often
- Dizziness

## Diagnosis

There is no laboratory test or imaging study that can detect fibromyalgia. The diagnosis is based on the patient's history and a physical examination. In 1990 the American College of Rheumatology (ACR) specified that the patient must have widespread pain for at least three months and soreness at eleven out of eighteen specific pressure points. These points are located on the neck, shoulders, chest, rib cage, lower back, thighs, knees, elbows, and buttocks. Diagrams of these pressure points are available on the ACR website at http://www.rheumatology.org/public/ factsheets/fibromya\_new.asp?aud=pat.

## Treatment

Treatment of fibromyalgia is usually broadbased, consisting of recommendations about exercise and regular sleeping habits as well as medications. A variety of different drugs have been prescribed for fibromyalgia, including

## What Is Pain?

There is some evidence that fibromyalgia is associated with increased sensitivity to pain in some patients. Pain is, however, more than a physical process. Studies of the brain have shown that people's perception of pain has emotional and cognitive (thought-related) dimensions as well. In other words, a person's feelings and thoughts about pain have some influence on how intensely they experience pain. Some people grow up in families in which their relatives frequently complain about pain, while others are taught to ignore physical discomfort. Because of the different ways that people learn to cope with pain as they are growing up, some doctors think that people with fibromyalgia are focusing too much on aches and pains that others take in stride as part of life.

The relationship between thoughts and feelings on the one hand and perception of physical pain on the other explains why patient education and stress management programs are often recommended for people with fibromyalgia. People can learn to cope more effectively with the physical pain they experience and avoid reinforcing it by allowing it to control their lives. As one doctor explains it, health care professionals can help patients with fibromyalgia "to not choose illness as a way to deal with difficult life situations."

muscle relaxants, tranquilizers, antidepressants, and pain relievers; however, many doctors are cautious about these drugs because of the possibility that the patient will become dependent on them.

A new drug called Lyrica was approved by the FDA in the summer of 2007 as a treatment for fibromyalgia. The new drug was originally developed to treat seizures and nerve pain associated with diabetes. It is controversial as a treatment for fibromyalgia, however, because it has several potentially serious side effects, such as weight gain, blurred vision, swelling of tissue, sleepiness, and dizziness. In addition, Lyrica may not provide patients with long-lasting benefits, as people with fibromyalgia frequently change from one drug to another every few months.

In some cases the patient's doctor may recommend psychotherapy to help the patient regain a sense of control over his or her life and to learn better ways to cope with physical pain. Relaxation techniques may be useful.

Some complementary and alternative treatments appear to be beneficial to people with fibromyalgia. These include therapeutic massage, acupuncture, and chiropractic. It is better, however, for patients to use approaches that get them involved in their own healing, such as regular exercise, losing weight if they are too heavy, and endurance and strength training, rather than therapies in which they are simply receiving treatment from another person.

### Prognosis

Fibromyalgia can last for years but is rarely fatal. The patient's symptoms may get better on occasion and then worsen again during stressful periods; one survey found that fibromyalgia patients average ten outpatient visits to their doctor every year and are hospitalized once every three years.

### Prevention

There is no known way to prevent fibromyalgia.

### The Future

It is uncertain whether fibromyalgia will be diagnosed more frequently in the future. Since the disorder was identified only a few decades ago, it is possible that it will be redefined at some point, along with other similar pain and chronic fatigue syndromes.

**SEE ALSO** Chronic fatigue syndrome; Gulf War syndrome; Irritable bowel syndrome; Lyme disease; Restless legs syndrome; Rheumatoid arthritis

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## WORDS TO KNOW

- Acupuncture: A form of alternative medicine in which very fine needles are inserted into the skin at specific points on the body for pain relief.
- **Chiropractic:** A form of alternative medicine that treats disorders of the joints and muscles by adjusting the patient's spine or other joints.
- **Fibro fog:** A term that has been coined to describe memory loss and difficulty concentrating in fibromyalgia patients.
- **Pressure points:** Specific locations on the body where people with fibromyalgia feel pain even with light pressure.
- **Rheumatology:** The branch of medicine that deals with disorders of the muscles, joints, and connective tissue.
- **Substance P:** A chemical in the central nervous system that transmits pain signals back and forth between the brain and the rest of the body.

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# Flesh-eating disease

See Necrotizing fasciitis.



# Food Poisoning

## Definition

Food poisoning is a general term for any illness of the digestive system that results from eating food. The food may be contaminated by bacteria, viruses, parasites, chemicals, or natural toxins produced by certain plants or animals.

## Description

Food poisoning is usually a disorder of the stomach and intestines resulting from contaminated food. The contamination may result from not cooking meat and other animal products thoroughly; from washing food in contaminated water; from failing to refrigerate leftover food promptly; or from food being handed by people who are carrying disease organisms. Some forms of food poisoning affect other parts of the body, especially the nervous system and/or kidneys.

## **Demographics**

According to the Centers for Disease Control and Prevention (CDC), there are on average 76 million cases of food poisoning in the United States each year; most are mild illnesses that run their course in a few days with appropriate self-treatment at home. About 325,000 people, however, become sick enough to require hospital treatment, and 5,000 will die. Those at greatest risk of serious illness or death from food poisoning are the very young, the very old, people with weakened immune

### Also Known As

Foodborne illness, acute gastroenteritis

### Cause

Bacteria, bacterial toxins, viruses, parasites, chemicals, toxic plants or animals

### Symptoms

Nausea, vomiting, diarrhea, headache, weakness, abdominal cramps

**Duration** One to ten days systems, and people exposed to an unusually high dose of a disease organism.

The profile of foodborne diseases has changed over the years. At the turn of the twentieth century, such diseases as cholera, typhoid fever, and tuberculosis were the most common illnesses caused by contaminated food or water. The pasteurization of milk, safer methods of canning food, and better purification of drinking water have made these diseases increasingly rare in the developed world. Newer diseases have emerged, however, because bacteria mutate and evolve over time. In addition, air



*Image of* Salmonella *bacteria*. © MEDICAL-ON-LINE / ALAMY.

travel and other methods of rapid transportation mean that disease organisms that used to be limited to one part of the world can spread to other countries in days rather than years.

### **Causes and Symptoms**

Food poisoning can be caused by a wide range of microorganisms, chemicals, and natural toxins. About 60 percent of cases of food poisoning in the United States and Canada are caused by bacteria that live in the intestines of chickens, turkey, and cattle, or by toxins produced by these bacteria; another 30 percent are caused by viruses. Food poisoning from chemicals or naturally poisonous plants or animals is less common.

- Bacterial infections. These are the most common type of foodborne illness in Europe and North America; 97 percent of these cases are related to improper food handling, 79 percent from food prepared in restaurants or cafeterias and 21 percent from improper cooking or storage of food at home.
- Parasites. These include tapeworms, flatworms, and amoebas, often found in raw fish or contaminated drinking water.
- Viruses. About 30 percent of cases of food poisoning in the United States result from eating food contaminated by Norwalk virus, rotaviruses, and certain forms of the hepatitis virus.
- Chemicals. Most cases of chemical contamination of food in the United States are caused by pesticide residue on fruits or vegetables, but there are also cases of people adding chemicals directly to food in the belief that they were using ordinary table salt.

## A Treat to Die For

Most people would not think of eating an animal known to be poisonous as part of a gourmet meal in a fancy restaurant. For generations, however, the Japanese have considered fugu, or puffer fish, a great delicacy when the fish's flesh is sliced thin and served in an attractive pattern on a plate. Fugu can cost as much as \$200.00 in one of the better restaurants in Tokyo. Unfortunately, the liver, skin, and intestines of the puffer fish contain a deadly nerve poison that has no antidote. The Japanese still remember a famous actor who ordered a serving of fugu liver one night in 1975 because he enjoyed the tingling sensation as it passed his tongue and lips. Within minutes, however, his arms and legs were

paralyzed, then his breathing muscles. Eight hours later he died.

Although fugu is still served in Japan, chefs who prepare it must study for two years and pass a stiff examination to show that they can separate the fish into poisonous and nonpoisonous parts as well as prepare it for customers; only about 30 percent of the chefs succeed in passing the examination. More recently, scientists at Nagasaki University have reported success in breeding a nontoxic variety of puffer fish by restricting the fish's diet. According to the scientists, it is the food that the fish eats in the wild plus its digestive process that make its liver and intestines so poisonous to humans.



Fugu fish in Japan. SHUTTERSTOCK.

 Natural toxins. Natural toxins are substances produced by certain mushrooms and plants, and several species of reef fish that are poisonous to humans by ingesting toxins produced by algae that come through the food chain.

The most common symptoms of food poisoning are nausea, vomiting, abdominal cramping, and diarrhea. In the case of food poisoning caused by bacteria and viruses, the organisms either directly irritate the tissues that line the intestines or produce toxins that destroy the cells of the intestinal lining. The digestive tract secretes large quantities of fluid in order to flush the invaders out of the body, resulting in loose, frequent, and watery stools. Vomiting is caused by bacterial toxins acting on the central nervous system.

The timing of the symptoms varies, depending on the specific organism or natural toxin involved, the amount of food that the person ate, and the person's overall health. People can feel sick within a few hours, even a few minutes, after eating contaminated food, or it may take several days before they feel ill. Symptoms caused by parasites may not appear for two weeks or longer.

## Diagnosis

Many people do not seek medical help for a mild case of food poisoning. If a person does go to the doctor, the doctor will begin by taking a history of the person's recent eating habits, including where, as well as what, they ate. Because food poisoning can occur as a group outbreak as well as affecting individuals, the doctor will want to know whether other people might be sick and whether the public health department should be notified.

In most cases the doctor will treat the patient for diarrhea and nausea without trying to determine the specific organism that contaminated the food, because laboratory tests for viruses or bacteria require specialized equipment and techniques. If the doctor thinks that it is important to identify the organism, he or she can take a sample of the patient's stool for laboratory analysis. This method can also be used to detect and identify parasites. In some cases the doctor may take a blood test if the patient has a high fever or other signs of an acute infection.

If the patient's food history indicates that they ate a poisonous plant or animal, they will usually be taken to a hospital as quickly as possible for specialized treatment.

### Treatment

Treatment consists of rest and preventing dehydration. If the patient has lost a lot of body fluid from repeated episodes of diarrhea or vomiting, they are encouraged to drink clear liquids with small amounts of salt and sugar added. A solution to replace fluid can be made at home by adding one level teaspoon of salt and four heaping teaspoons of sugar to one quart of water. People who are severely dehydrated may need to be taken to the hospital and given fluids intravenously.

The doctor may also prescribe medications to stop the diarrhea, such as Pepto-Bismol or Kaopectate. If the patient's diarrhea lasts more than four days, the doctor will order a stool culture (sample to be sent to a laboratory to be tested) to identify the organism. Other signs that further treatment is needed are: fever higher than 101°F (38.3°C), blood in the stools, or prolonged vomiting and severe dehydration. Depending on the organism, the doctor may prescribe an antibiotic.

### Prognosis

Most people recover completely from food poisoning in a few days. Complications are rare except in infants, the very old, and people with HIV infection or other diseases that weaken the immune system.

### Prevention

People can reduce their risk of food poisoning by observing simple precautions recommended by the CDC when cooking at home:

- Cook meat, eggs, poultry, fish, and other animal products thoroughly. Ground beef should be heated to an internal temperature of 160°F (71°C), poultry to 180°F (82°C), and fish to 140°F (60°C).
- Separate foods. Do not cut fruits and vegetables on a cutting board that has been used for raw chicken or beef until the cutting board has been washed. Do not put cooked meat on a plate that has held raw meat.
- Chill leftovers promptly. Cooked food should be refrigerated within four hours after a meal to prevent bacteria from growing in it. Keep the refrigerator temperature at 40°F (4°C).
- Clean fruits and vegetables in running water and remove the outermost leaves of a head of cabbage or lettuce.
- Wash hands before and after food preparation; do not prepare food for others when ill.

## WORDS TO KNOW

- **Carrier:** A person who has an infectious disease and can transmit it to others but has no symptoms of the disease themselves.
- **Dehydration:** Loss of water from the body. It may be caused by fever, vomiting, diarrhea, or excessive sweating.
- **Irradiation:** A technique for treating raw meat and poultry with gamma rays, x rays, or electron beams to destroy disease organisms.
- **Pasteurization:** A process in which milk or fruit juice is partially sterilized by heating to a temperature that destroys disease bacteria without causing major changes in appearance and taste.
- **Toxin:** A poisonous substance produced by a living cell or organism.

• Report suspected foodborne illnesses to the local public health department; this includes foods purchased in supermarkets as well as eaten in restaurants.

Tips for food safety when traveling:

- Do not eat wild plants or mushrooms when hiking or camping.
- Drink only boiled water when traveling abroad, and avoid ice, salad, and raw fruits, vegetables and seafood. Eat only hot, freshly cooked food.
- Consult the CDC for travelers' advisories about specific countries.

## The Future

New technologies may help to reduce the risk of food poisoning in the future by making foods safer. One technique that has already proven useful is irradiation of ground beef and other meats; another is a new way to pasteurize raw eggs in the shell. Another promising field of research is to study the ways in which disease organisms are spread among animals. Finding ways to prevent animals from becoming infected by bacteria and viruses would lower the rate of food poisoning among humans.

### **SEE ALSO** Toxoplasmosis

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## **Fractures**

### Definition

A fracture is a break in a bone. It may extend only partway through the bone (called a partial fracture) or be a complete break that goes the whole way across the bone.

Fractures can be classified in several different ways. They may be categorized by the body part or specific bone that is broken, or by certain characteristics of the fracture:

- Simple fracture: The bone is broken in only one place.
- Multi-fragmentary (also called comminuted) fracture: The bone is broken in several places and there are at least three bone fragments.
- Closed fracture: The skin over the injury is not broken.

Also Known As Broken bones

### Cause

Trauma (falls, sports injuries, accidents); weakened bones

### Symptoms

Pain; inability to move injured part; protruding bone, bleeding, or swelling at injury site

### Duration

Three or four weeks to several months

- Open (also called compound) fracture: The skin has been cut open over the fracture. The bone may or may not be visible through the wound.
- Greenstick fracture: The bone is broken on one side while the other side is bent. This type of fracture takes its name from the typical breakage pattern of a green tree limb.
- Undisplaced fracture: The pieces of the broken bone are aligned.
- Displaced fracture: The pieces of the broken bone are not in alignment.
- Stress fracture (also called hairline fracture): A thin partial break in a bone caused by repeated stress on the body part. Stress fractures are most likely to occur in athletes.
- Compression fracture: A fracture that occurs in the spine of a person with osteoporosis. The front part of a vertebra collapses because the bone has been weakened by loss of calcium.



## Description

The experience of a fracture varies considerably depending on the location of the bone that has broken, the person's age, the cause of the fracture, and the amount of force that was applied to the bone. Most broken bones are caused by a sudden sharp blow, fall, gunshot wound, or other trauma, and the person often hears a snapping or grinding noise as the bone breaks. Stress and compression fractures, however, result from long-term overuse or weakening of the bone, and develop gradually.

Children's bones are more flexible than those of adolescents or adults, and greenstick fractures are more common in their age group. Adults are more likely to break the bone completely. Elderly people are more likely to develop compression fractures; they are also more likely to break bones as the result of falls, and to suffer broken bones from less force than it would take to cause a broken bone in a younger person. *X ray showing the fracture of two leg bones in a ten-year-old boy.* © PHOTOTAKE INC. / ALAMY.

## **Basic First Aid for Fractures**

The seriousness of a fracture depends partly on its location. A broken back or neck, or head injury, is a medical emergency. If an injured person has a broken neck or back, or if they have a broken bone that is coming through the skin:

- Call for emergency help at once.
- Do *not* try to move the injured person. Wait for help to arrive.
- If there is bone coming through the wound, have the person lie down. Apply pressure to the wound with a gauze pad or clean cloth, but do *not* try to push the bone back through the skin or wash the wound.

If the fracture is less serious:

- Remove clothing from the injured limb. Do *not* try to force an arm or leg backward through a sleeve or pants leg; cut the garment away with scissors.
- Apply a cold compress or ice wrapped in cloth to the injured part.
- Make a temporary splint. Keep the injured limb in the position it is in at the time of injury; then place a towel or other soft padding around the injured part. Then take a board or other firm object and place it next to the injured part outside the padding. Make sure that the board is long enough to cover the joints above and below the fracture. Use first-aid tape to hold the splint in place.
- Do not give the injured person anything to eat, in case they must be taken into surgery when help arrives.

## Demographics

Fractures are a common injury in the general American population, although the location of fractures varies somewhat by age group. The average American can expect to suffer two fractures over the course of his or her life. According to the National Center for Health Statistics, there are an average of 1.1 million hospitalizations and 3,450,000 visits to outpatient emergency departments each year for fractures. An additional 2 million fractures are treated in doctors' offices. Some specific annual statistics follow:

- Hip fractures: About 320,000.
- Fractures of the tibia (long bone in the lower leg): About 600,000; most caused by high-speed accidents, with the highest rates in teenaged males.
- Collarbone fractures: About 400,000.
- Stress fractures: More common in women than in men; found in 45 percent of competitive female long-distance runners.
- Fractures of the lower neck: About 11,000.

In people under age seventy-five, the most common fracture is a wrist fracture. In people over seventy-five, hip fractures are the most common injury. In people younger than fortyfive, men are more likely to suffer fractures than women; in adults over forty-five, however, women are more likely than men to suffer broken bones. Over age sixty-five, women are three times as likely as men to break a bone.

More than two-fifths of fractures in the United States occur at the person's home, 23 percent inside the house, and 19 percent on the grounds outside the house.

## **Causes and Symptoms**

Most fractures result from some kind of trauma, most often a fall, highimpact blow, or other accident. Athletes can develop stress fractures from overexercise, however, and people with osteoporosis can develop compression fractures from the simple weight of the spinal column on the lower back with little or no trauma.

With the exception of stress and compression fractures, people can often hear a bone snap or break when it fractures. The classic symptoms of a fracture are:

- Intense pain
- Affected body part looks misshapen or out of place
- Numbness or a tingling sensation
- Swelling, bruising, or bleeding of surrounding skin
- Difficulty in moving affected part or complete inability to move it

In stress fractures, the patient usually has no history of trauma but complains of pain in the affected part of the body that has come on gradually and gets worse during activity.

### Diagnosis

In most cases the patient's history is the key to the diagnosis, including age, sex, occupation, sports participation, and overall health as well as a recent fall or other accident.

The doctor will order an x ray to determine the location and pattern of the fracture. In the case of stress fractures, the doctor will look for swelling or redness in the affected body part, and press gently on the tissues around the bone for signs of increased pain or tenderness.

### Treatment

Normal bone heals itself following a fracture. Blood flows into the area of a break and forms a clot. The clot is gradually replaced by a matrix of collagen fibers. Cells called fibroblasts begin to lay down lines of new bone cells on both sides of the fracture line. As these lines of cells grow toward each other, the break in the bone is gradually repaired. Treatment of a broken bone is therefore focused on holding the broken bone in the proper position while the break is healing. The bone may be held in place by either external or internal fixation methods. External fixation methods include casts, splints, or cast-braces. These are usually made from fiberglass or plaster. Internal fixation methods include pins, screws, or wires inserted by a surgeon to hold larger bones in place. Surgery may also be required to smooth the edges of broken bones or to remove bone fragments in order to prevent infection. Open (compound) fractures require the administration of antibiotics and careful antiseptic cleansing of the wound to prevent infection.

Most fractures require about four weeks to heal. While some use of the injured body part speeds healing, too much activity too soon can cause further damage. Since the pain of a fracture usually stops long before the bone is strong enough to return to normal levels of activity, the patient should limit the use of the injured part according to the doctor's advice. Follow-up x rays are usually ordered to make sure that the bone is in proper alignment and that the fracture is healing normally.

Many people require some form of physical therapy or rehabilitation after the cast or other fixation materials are removed because the muscles have grown weak from lack of exercise and the ligaments near the healing bone may feel tight or stiff.

### Prognosis

The prognosis of a fracture depends on its cause and location, the patient's age and overall health, and the severity of the fracture. Most fractures will heal eventually with prompt treatment. The patients at highest risk of lasting complications from fractures are those with osteoporosis or other diseases of the bone; those whose fractures involved crushing as well as breaking of the bone; those with compound fractures that became infected; those who are heavy smokers; those with poor nutrition; and those over sixty years of age. Elderly people with hip fractures are 5–20 percent more likely to die within a year of their injury than others in their age group.

### Prevention

Children, adolescents, and young adults can reduce their risk of fractures by:

- Wearing proper protective gear for athletic activities and following appropriate safety measures.
- Eating a nutritious diet rich in calcium.

## WORDS TO KNOW

**Fibroblast:** A type of cell that provides structure during the healing of a broken bone or other wound.

**Vertebra:** One of the 33 bones that make up the spinal column in humans.

- **Fixation:** The medical term for holding a broken bone in its correct position to speed healing and prevent further injury.
  - Avoiding overexercising and overly restrictive diets. Many female athletes develop stress fractures as a result of trying to lose more weight than is healthy for them.
  - Not smoking. Smoking slows down the process of bone healing after a fracture.
  - Wearing properly fitting shoes, and replacing athletic shoes as soon as the soles begin to wear unevenly.

Elderly people need to take special precautions because of the longterm dangers of fractures resulting from falls:

- A safety inspection of the house or apartment, checking for slippery floors, the lack of hand rails on stairs or in the bathroom, poor lighting, loose rugs, and other features that may increase the risk of falls.
- Asking the doctor whether any medications that are prescribed have drowsiness or loss of coordination as side effects.
- Being very careful about alcohol intake. Alcohol can not only make people dizzy or lose their balance but also encourage risky behaviors that increase the risk of a fall.
- Wearing shoes with low heels that fit properly, and avoiding walking around the home in stocking feet or loose slippers.
- Keeping bones strong by getting enough calcium and vitamin D, and exercising regularly. Women should have a bone density test every few years according to their doctor's recommendations.
- Practicing balance exercises at home to improve flexibility as well as balance.

• Asking the doctor about hip padding. Some doctors recommend it as a way to lower the risk of a hip fracture if the person does fall.

### The Future

Fractures are likely to be a commonplace injury for people of all ages, in younger patients as a result of sports or outdoor-related activities and car or motorcycle accidents, and in elderly people as a result of falls. As the population of older adults continues to grow, the number of fractures in this age group is likely to increase as well.

SEE ALSO Osteoporosis; Sprains and strains

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# Fragile X

## Definition

Fragile X syndrome is a genetic disorder caused by a mutation in the FMR1 gene on the X chromosome. This gene makes a protein called fragile X mental retardation 1 protein, whose function is not fully understood but is thought to be related to the development of synapses, which are specialized junctions between nerve cells. The mutation in the gene blocks production of the needed protein, leading to mental retardation and other developmental problems in affected children. The disorder is called fragile X because the mutated gene causes part of the chromosome to look unusually slender or fragile under a microscope.

## Description

Fragile X syndrome is a genetic disorder caused by a mutation in the FMR1 gene on the X chromosome. The disorder was first described in 1943 by two psychiatrists named Martin and Bell, who determined that the syndrome is sex-linked. The specific gene involved, however, was not identified until 1991.

The mutation consists of the expansion of a section of the gene that ordinarily consists of six to fifty-five repeats of a specific trinucleotide (tripletnucleotide; nucleotides are one of the building blocks of DNA). In some people, this portion of the FMR1 gene contains more than fifty-five repeats of the triplet. People with sixty to 200 repeats have what is called a premutation; they do not have the typical symptoms of fragile X syndrome but they can carry the defective gene and pass it on to their children. The repeated portion of the FMR1 gene is likely to have more repetitions added when it is passed from a woman with the premutation to her children. When the number of repetitions grows to about 230, the gene is switched off and does not produce the protein that it normally makes. This gene change is then considered a full mutation. Some people with fragile X syndrome have as many as 1,000 repetitions of the crucial triplet in the FMR1 gene.

Because the X chromosome is a sex-linked chromosome, fragile X syndrome affects boys more severely than girls. Girls have two X chromosomes, whereas boys have one X chromosome and one Y chromosome. A boy who inherits a full mutation of the FMR1 gene will develop fragile X

### Also Known As

Marker X syndrome, Martin-Bell syndrome, Escalante's syndrome

### Cause

Mutation in the FMR1 gene

### Symptoms

Mental retardation, elongated face, hyperactivity, anxiety, autistic behavior, seizures

### Duration Lifelong



Although fragile X syndrome causes some mental handicaps, boys like Zach Weaver (right) can still participate in most activities. AP IMAGES.

syndrome because his only X chromosome contains the mutated gene. A girl who inherits a full mutation may not be as severely affected because each cell of her body needs to make use of only one of its two X chromosomes; it can inactivate the other X chromosome.

People with a fragile X premutation are usually of normal intelligence but may suffer from other health problems. About 20 percent of women with the premutation stop having menstrual periods unusually early, often by age forty. Men and some women with the premutation are at increased risk of developing a movement disorder accompanied by memory loss, tremor, and loss of sensation in the lower legs.

## **Demographics**

Fragile X syndrome is the most common genetic cause of mental retardation in boys, affecting one in 4,000 males and one in 8,000 females. A screening study that was done of children in special education programs in the United States found that approximately

one in 400 males receiving special education services have fragile X syndrome. According to the Centers for Disease Control and Prevention (CDC), about one in 259 women carry the fragile X mutation and could pass it to their children. About one in 800 men carry fragile X changes; their daughters will also be carriers. As far as is known, fragile X syndrome is equally common in all races and ethnic groups; it appears to be one of the most common genetic disorders in humans. No large-scale population screening in any country in the world has been carried out, however.

## **Causes and Symptoms**

The cause of fragile X syndrome is a mutation in the FMR1 gene on the X chromosome. Not all children are affected to the same extent by the mutation, however. Girls who inherit the fragile X may appear normal or they may have some degree of mental retardation, but usually to a

lesser degree than boys with the syndrome. Between 33 and 50 percent of girls with fragile X syndrome have significant intellectual impairment; the rest have either normal intelligence or specific learning disabilities, such as mathematics. Emotional and behavioral problems occur in children of either sex. Children with fragile X syndrome have characteristic physical features as well as developmental and behavioral difficulties:

Physical features:

- Elongated face with large ears
- Flat feet
- Double-jointed fingers and easily dislocated joints
- Poor muscle tone
- Excessive curvature of the spine
- In males, unusually large testicles

Mental symptoms:

- Mental retardation (IQ between 20 and 70)
- Difficulty with speech and language development

Behavioral symptoms:

- Hyperactivity
- Some autistic behaviors (poor eye contact, extreme shyness, and hand flapping)
- Problems paying attention
- Aggressiveness
- About 20 percent of boys with fragile X meet all the diagnostic criteria for autism. Most boys and some girls with fragile X have some symptoms of autism; however, many are socially outgoing and can form relationships with other people.

Neurological symptoms:

- Anxiety
- Depression
- Rapidly changing moods
- Seizures (about 25 percent)

## Diagnosis

The diagnosis of fragile X syndrome is based on a combination of family history, early signs of mental retardation in the child, and specific genetic

testing. Patients who have several mentally retarded male relatives or a mother with mental retardation or learning disabilities are sometimes screened on the basis of this family history. In other cases the child may not be diagnosed until he or she has started school and his or her learning disabilities become apparent. The standard diagnostic test for fragile X syndrome is a DNA test that was developed in 1992. It counts the number of repetitions in the FMR1 gene and can detect carriers of the mutation as well as fully affected persons. The doctor takes a sample of the patient's blood and sends it to a laboratory for analysis. The test is expensive but is often covered by health insurance.

### Treatment

There is no cure for fragile X syndrome. Treatment usually involves a combination of approaches. The syndrome can be treated through cognitive behavioral therapy, special education programs, speech and language therapy, medications for anxiety and depression, and treatment of physical abnormalities if needed. Because fragile X syndrome is not a rare condition, specific educational approaches for these children have been developed and tested. Most children affected by the syndrome, however, will need some form of treatment throughout their lives.

### Prognosis

The prognosis for a child with fragile X syndrome depends on sex and on the number of repeated triplets in the FMR1 gene. Girls usually have a better prognosis for intellectual development than boys; children with fewer repeats in the gene have a better prognosis than those with several hundred. People with fragile X syndrome do, however, have a normal life expectancy.

### Prevention

There is no way to prevent the genetic mutation that causes fragile X syndrome. Parents who have a child with the condition should consult a genetic counselor, however, to determine the risk of having another child with the syndrome or a child who is a carrier of the mutation. Accurate diagnosis is important because of the possibility of other family members inheriting either the syndrome itself or other problems related to an increased number of repeats in FMR1.

## WORDS TO KNOW

- **Nucleotides:** The basic structural units of DNA and RNA.
- **Premutation:** An abnormally large number of repeated triplets in certain genes that does not cause obvious symptoms of a genetic disorder but can expand into a full mutation when transmitted to offspring.
- **Synapse:** The medical term for specialized connections between nerve cells.
- **Triplet:** In genetics, a unit of three nucleotides that starts or stops the production of a specific protein. Triplets are also called codons.

## The Future

Research is focused on learning more about the FMR protein. It is known that the lack of the protein does not destroy nerve cells but rather delays their development. It is possible that a treatment can be found that will help the nerve cells and the synapses that join them to develop normally.

**SEE ALSO** Attention-deficit hyperactivity disorder; Autism; Depression; Down syndrome

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# Frostbite

## Definition

Frostbite is a disorder of the skin and underlying tissues caused by exposure to extreme cold and freezing of tissue.

## Description

Frostbite is most likely to affect the face, hands, and feet; however, the shins, knees, and the outer portions of the eyes may also be affected. Freezing of exposed tissues results in the formation of ice crystals inside the cell wall.

## Demographics

Frostbite is most likely to occur among soldiers, people who work outdoors in cold weather, mountain climbers, skiers and other winter sports participants, homeless people, travelers stranded outside in cold weather, and people who live close to the polar regions. In a few cases frostbite is caused by industrial accidents, when workers who must handle liquid nitrogen or other liquefied gases fail to protect their hands or use proper safety equipment. It is estimated that frostbite in North America and northern Europe causes 2.5 hospital admissions per 100,000 people. The true rate is unknown because there is no standardized reporting system for this disorder.

Most frostbite victims are male, but this ratio is thought to reflect occupational choices and interest in outdoor sports rather than a genetic factor.

### Also Known As

Cold exposure to the extremities, cold-related injury

### Cause

Exposure to extreme cold

### Symptoms

Numbness or pain, reddening or swelling, increased sensitivity to cold, clumsiness, sore joints

### Duration

One to three months; amputation is a lifelong injury

Seaborn Beck Weathers (left) suffered severe frostbite while attempting to climb Mount Everest. AP IMAGES.



According to U.S. military statistics, African American male soldiers are four times as likely and African American female soldiers 2.2 times as likely to suffer frostbite as their Caucasian counterparts. Other ethnic and racial groups from warmer climates are also thought to be more likely to suffer frostbite. In addition to race, certain diseases, including diabetes, thyroid disorders, arthritis, and some infections increase a person's risk of developing frostbite during exposure to cold.

Most frostbite victims are middle-aged adults between the ages of thirty-five and fifty; one study found the average age of patients treated for frostbite was forty-one.

## **Causes and Symptoms**

Frostbite is caused by exposure of skin and underlying tissues to extreme cold. When the skin is exposed to temperatures at or below 32°F (0°C), the blood vessels in the skin start to constrict. This closing down of the

# The High Cost of Mountain Climbing

Many famous mountain climbers have lost fingers or toes to frostbite on their expeditions. One of the most tragic cases involved Maurice Herzog (1919–), a French climber who was the first to climb Annapurna, the tenth-highest mountain in the world. Herzog and his fellow climber, Louis Lachenal (1921-1955), lost all of their toes and Herzog most of his fingers as a result of frostbite. The two men had chosen boots that were too thin in order to reach the summit of the mountain as guickly as possible, and Herzog lost his gloves near the summit in a freak accident. The gangrene suffered by both men meant that the expedition's doctor had to amputate their fingers and toes in the field without benefit of anesthesia.

Willi Unsoeld (1926–1979) was an American climber who was famous for getting to the top of Mount Everest in 1963 by a previously untried route even though he was not the first person to reach the summit. Unsoeld developed frostbite in his feet on the way down from the summit of Everest; he lost all his toes and had to spend several months in the hospital. Unsoeld later taught courses at Evergreen State College in Washington. He died in an avalanche while climbing Mount Rainier in March 1979. blood flow in the extremities is the body's protective strategy for preserving normal body temperature in the body core (the heart and other internal organs).

The early stage of frostbite is sometimes called frostnip. Short-term symptoms include loss of feeling or aching pain in the affected part, followed by redness of the skin and tissue swelling. Long-term symptoms include intense pain in the affected part, tingling sensations, cracks in the skin, dry skin, loss of fingernails, joint stiffness, loss of bone or muscle tissue, and increased sensitivity to cold. If left untreated, frostbitten skin gradually darkens and blisters after a few hours. Skin destroyed by frostbite is completely black, looks burned, and may hang loosely from the underlying tissues.

## Diagnosis

Diagnosis is usually made in the field on the basis of the appearance of the frostbitten parts of the body. Some doctors use a four-degree classification of injuries:

- First-degree: The epidermis (outermost layer of the skin) is reddened, swollen, and may look waxy. There is also a loss of sensation in the affected skin.
- Second-degree: The skin is reddened, swollen, and has formed blisters filled with a clear or milky fluid.
- Third-degree: The blisters are filled with blood and the skin begins to turn black.
- Fourth-degree: The epidermis, dermis, and underlying muscles, tendons, and bones are damaged.

A technique that can be used to diagnose the extent of soft-tissue injury after frostbite is technetium scintigraphy. This is a technique in which radioactive technetium is administered intravenously. The radioactive element is taken up differently by healthy and damaged tissue, and the pattern of "hot spots" and "cold spots" as traced by a scanner allows the doctor to tell whether and where deep tissues have been damaged by frostbite. Scintigraphy can also be used to monitor the recovery of the injured tissues following emergency treatment.

X rays and other imaging studies will not help in diagnosing frostbite but may be used to evaluate the injured person for broken or fractured bones.

### Treatment

Treatment at the scene begins with treating any life-threatening conditions first (internal injuries, etc.). The injured person should have wet clothing replaced with soft dry clothing to prevent further heat loss. Rewarming of the injured part should not be attempted if there is any danger of refreezing; in addition, the injured areas should not be rubbed with either snow or warm hands, as such rubbing may make the injury worse by pushing the ice crystals in the frozen skin through the cell wall. Last, the injured person should not be given alcohol or tranquilizers, as these will increase loss of body heat.

The injured person should then be taken to a hospital emergency room as soon as possible. Treatment begins with fluid replacement to speed up blood flow to the frostbitten tissues. The person will then be rewarmed either with warm wet packs or with a whirlpool bath at 104–108°F (40–42°C). Dry heat is not used for rewarming. The patient may be given morphine for pain. Thawing of frozen tissue takes about twenty to forty minutes, after which the injured part is placed on a sterile sheet, raised, and splinted. Blisters filled with clear fluid can be debrided (surgically removed), but blood-filled blisters are allowed to remain in order to prevent infection.

If frostbitten skin is not treated and its blood vessels are affected, gangrene may set in. Gangrene is the death of soft tissue due to loss of blood supply. It may be treated by surgical removal of the affected tissue if caught early; otherwise, the surgeon may have to amputate the affected digit or limb in part to prevent bacterial infections from spreading from the dead tissue to the rest of the body. People with frostbite may be given penicillin or other antibiotics in the emergency room to prevent infection of the damaged tissue, and ibuprofen to treat inflammation.

It may take from one to three months for the frostbitten tissue to heal. In most cases, amputation can be delayed for that length of time to see whether the affected body part will recover.

### Prognosis

Patients with early recovery of sensation in the affected part, blisters filled with clear fluid, and healthy-appearing skin color have a better prognosis for full recovery than those whose skin has turned bluish, has blood-filled blisters, and looks frozen.

People who have recovered from frostbite have an increased risk of another episode during future exposures to cold. They should take extra precautions to dress properly for extreme cold or avoid it altogether. They may also notice that the frostbitten parts of their body are more sensitive to ordinary cold weather, and ache or tingle whenever they are outdoors.

About 65 percent of people with severe frostbite will eventually develop arthritis in the affected hand, foot, or leg.

### Prevention

Frostbite can be prevented by wearing suitable clothing for weather conditions; wise preparation for dressing appropriately includes obtaining a weather forecast before going outdoors for work or recreation in cold weather. The head, neck, and face should be kept covered, and clothing should be loosely fitted and layered rather than tight-fitting; mittens are better than gloves for keeping hands warm. Hands and feet should be kept dry.

Other precautions include increasing calorie and fluid intake when outdoors in cold weather and avoiding the use of alcohol and tobacco. People who are hiking or skiing should use a buddy system in case one person is injured and must be evacuated quickly.

### The Future

It is difficult to predict whether the incidence of frostbite will increase in the future. Better patient education and more accurate long-term weather forecasting may help; however, many victims of frostbite are alcoholics, drug abusers, and mentally ill homeless people who are unaware of the dangers of exposure to cold or are too intoxicated to move indoors.

Some experimental treatments are being investigated to improve the flow of blood to frostbitten tissue or to treat the inflammation caused by frostbite; however, none of these treatments have been shown to be effective.

## WORDS TO KNOW

Amputation: Surgical removal of a limb.

**Debridement:** The medical term for the surgical removal of dead or damaged soft tissue.

Dermis: The layer of skin just below the epidermis.

Epidermis: The outermost layer of the skin.

**Gangrene:** Decay and death of soft tissue due to loss of blood supply.

**Scintigraphy:** A technique for detecting the location and extent of soft-tissue injury by injecting a small quantity of a radioactive element and following its distribution in the tissue with a scanner.

### SEE ALSO Gangrene; Hypothermia

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# G



### Cause

Loss of blood supply to soft tissue

### Symptoms

Numbness, blackened tissue, foul smell (skin); confusion, fever, severe pain (internal organs)

#### Duration

Hours (gas gangrene); days (wet gangrene); months to years (dry gangrene)



## Gangrene

### Definition

Gangrene is the medical term for the death of soft tissue as a result of the loss of blood supply to the affected part of the body. The loss of blood supply in turn can result from a disorder of the circulatory system, from trauma, or from infection. Gangrene most commonly involves the fingers, toes, hands, or feet, but can also develop within the internal organs.

There are two major types of gangrene, dry and wet. Dry gangrene is caused by a gradual loss of blood flow through the arteries. It typically develops slowly over a period of years as the person's circulation is impaired by atherosclerosis (hardening of the arteries), diabetes, or long-term smoking.

Wet gangrene develops much more rapidly than dry gangrene. It is caused by an untreated wound infection or by bacteria growing in tissue that has broken down as a result of pressure. Bedsores are a type of wet gangrene that may develop in patients who are bedridden for long periods of time.

Gas gangrene is a deadly subtype of wet gangrene caused by bacteria known as *Clostridium perfringens*. *C. perfringens* is an organism that grows in the absence of oxygen. It produces gas as well as toxins; these compounds expand and spread rapidly through nearby tissues. Gas gangrene is a medical emergency.

#### Gangrene

Gangrene of the foot. Severe gangrene can lead to amputation. © MEDICAL-ON-LINE / ALAMY.



### Description

Dry gangrene typically begins in the parts of the hands and feet that are furthest from the trunk of the body. These are the parts that are most likely to lose their blood supply in patients with diseases that affect the circulatory system. The affected area becomes cold and numb; it begins to turn red as the blood cells inside the smaller vessels begin to break down. It then turns a darker brown or blackish color. The dead tissue may eventually fall off if it is not removed by surgery first.

Wet gangrene develops when the blood flow to the affected body part is blocked—often by a traumatic injury—the blood that collects in the soft tissues supports the rapid growth of bacteria, which causes the tissues to swell and produce a foul-smelling discharge. The affected area is usually discolored and very painful, and the patient typically develops a fever. If the gangrene is not treated, the bacterial infection can spread throughout the body via the bloodstream and kill the patient. The possibility of death from spreading infection is the reason why military surgeons from the time of Augustus Caesar through the end of the nineteenth century routinely amputated the limbs of wounded soldiers; the operations were necessary to give the soldiers a chance to survive injuries to their arms or legs.

Gas gangrene is a type of wet gangrene caused by *Clostridium perfringens*, a bacterium that grows in the absence of oxygen. In addition to swelling and pain in the affected area, the tissue may also crackle or make a popping sound if the doctor presses on the affected area. The patient

## Death of a President

On September 5, 1901, President William McKinley (1843–1901) was shot by Leon Czolgosz at a reception at the Pan-American Exposition in Buffalo, New York. Czolgosz fired two bullets into McKinley's abdomen while waiting in line to shake hands with him. One bullet caused only a minor flesh wound, but the other passed completely through his stomach and pancreas and lodged in his back. McKinley was taken to the hospital and appeared to be improving following surgery. The surgeons did not, however, find the bullet as x-ray machines were still experimental in 1901. They closed up the wound with the bullet still inside.

McKinley continued to improve until September 12, when he developed the classic symptoms of gangrene caused by infection: headache, nausea, a rapid but weak pulse. He died two days later and was succeeded as president by Theodore Roosevelt.

A significant consequence of these events was a greater interest in the use of x-rays to detect the location of bullets inside the body. The official report of McKinley's autopsy noted that some organs along the track of the bullet through the president's abdomen had become gangrenous;



*President William McKinley.* COURTESY OF THE LIBRARY OF CONGRESS.

gangrene of the pancreas was listed as the cause of McKinley's death. Had modern x-ray equipment been available in 1901, McKinley would have almost certainly survived his wounds.

will become very sick very quickly with gas gangrene, often going into shock, sweating heavily, and developing mental symptoms (confusion and disorientation).

## **Demographics**

The frequency of gangrene depends on the type. Gas gangrene is fortunately rare in the United States, with an average of 900 to 1,000 cases reported each year. According to the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK), about 71,000 amputations of the feet and lower legs are performed each year in the United States on diabetics who have developed gangrene. More than 60 percent of all nontraumatic lower-limb amputations in the United States occur in people with diabetes.

Both wet and dry gangrene affect both sexes and all races equally. Risk factors for gangrene include:

- Old age. The elderly are more likely to develop circulatory disorders leading to gangrene or to be confined to bed and develop bedsores.
- Diabetes. Diabetes tends to weaken the blood vessels over time.
- Diseases of the blood vessels.
- HIV infection.
- Radiation or chemotherapy for cancer.
- Severe injury, including frostbite.
- Long-term smoking. The chemicals in tobacco smoke cause the blood vessels to narrow, reducing the supply of oxygen to body tissues.

### **Causes and Symptoms**

The basic cause of gangrene is the loss of blood supply to the soft tissues in a part of the body. This loss may result from circulatory disorders, from traumatic injury, or from infection.

The basic symptoms of wet and dry gangrene have already been described. Gangrene is a serious condition, and anyone with any of the following symptoms should see their doctor at once:

- Pain in the affected area that is out of proportion to the outward appearance of the wound.
- A foul-smelling ooze or discharge leaking from a wound or sore. In gas gangrene, the discharge has a mousy or slightly sweet odor.
- Skin that is colorless, pale, cold, and numb.
- Skin that is turning blue, brown, or black.

### Diagnosis

The diagnosis of gangrene is based on a combination of the patient's history, a physical examination, a blood test, and imaging studies. The doctor will ask the patient about any recent injuries, exposure to extreme cold, recent surgery, chronic diseases (particularly diabetes), smoking habits, and any disorders that affect the immune system. The physical examination will include examination of the affected body part as well as taking the patient's pulse, temperature, and blood pressure. Tests and imaging studies that are usually done to evaluate gangrene include.

- Blood test. A higher than normal white blood cell count usually indicates an infection. A blood test may also be done to identify the specific bacterium causing the infection (in wet gangrene).
- X-ray study. This test may be performed to look for gas bubbles in soft tissue when gas gangrene is suspected.
- Computed tomography (CT) scans and magnetic resonance imaging (MRI). These tests may be ordered to check for the spread of gangrene.
- Arteriogram. An arteriogram is a test in which a dye that shows up on an x-ray is injected into the patient's blood circulation. It can help the doctor determine whether any of the patient's arteries are blocked. This type of test is usually done for patients with dry gangrene.
- Removal of a sample of the affected tissue. The sample can be examined under the microscope for signs of tissue death.
- Removal of a sample of fluid from the affected area. The fluid can be cultured in a laboratory to look for signs of *C. perfringens* when gas gangrene is suspected.

In some cases the patient may be taken directly into surgery to determine how far the gangrene has spread inside the body.

### Treatment

Gangrene is a serious condition requiring treatment in a hospital. A patient diagnosed with dry gangrene will be assessed by a surgeon who specializes in vascular disorders to determine whether it is possible to restore blood flow to the affected area. The tissue that has already died, however, is usually removed surgically.

Wet gangrene requires immediate treatment with intravenous antibiotics to stop the spread of infection and debridement (surgical removal of the dead tissue). If the affected area is large, the patient may need a skin graft to restore the appearance of the limb. In severe cases, the patient's toes, fingers, or limbs may have to be amputated to prevent the spread of infection to vital organs.

Gas gangrene is a medical emergency. The patient is given intravenous antibiotics while the wound is debrided. Following surgery, the
patient may be placed in a hyperbaric oxygen chamber, a room in which pure oxygen under pressure is delivered to the patient. Hyperbaric oxygen raises the oxygen levels in the patient's blood, which in turn may help to fight the bacteria that cause gas gangrene.

#### Prognosis

The prognosis depends on the type of gangrene, the length of time before the patient sought treatment, and the patient's age, underlying medical conditions, and immune status. Dry gangrene usually has a better prognosis than wet gangrene because it does not involve infection. Gas gangrene has a mortality rate ranging from 25 percent in otherwise healthy patients to 67 percent in patients being treated for cancer.

## Prevention

All people can prevent gangrene by cleansing wounds promptly with an antiseptic; by watching for signs of infection in the wound (pus, redness, swelling, and unusual pain); and by going to the doctor at once if a wound looks infected.

People with diabetes need to take special care to prevent gangrene in the feet, which is a potential complication of the disease. The reason for this complication is that diabetes sometimes affects the nerves in the feet, causing a loss of sensation. The diabetic person may not notice a cut or other injury to the foot because they cannot feel it. An infection can then develop and lead to gangrene. NIDDK recommends that people with diabetes take the following precautions to prevent gangrene in their feet:

- Wash the feet in warm (not hot) water every day and dry them completely, particularly between the toes.
- Check the feet every day for cuts, blisters, swelling, redness, calluses, or toenail problems.
- Cut the toenails once a week after washing the feet.
- Always wear shoes or slippers to protect the feet from injuries.
- Always wear socks or stockings to prevent blisters, even when wearing sandals.
- Wear shoes that fit well, and check them before wearing to make sure there are no rough edges that might injure the feet.

# WORDS TO KNOW

Amputation: Surgical removal of a limb.

**Bedsore:** A type of wet gangrene that develops when a bedridden person cannot turn over to relieve pressure on soft tissue caused by the weight of the body. Bedsores are sometimes called pressure ulcers.

**Debridement:** The medical term for the surgical removal of dead or damaged soft tissue.

# The Future

Gangrene will always be a potential complication of traumatic injuries as well as such chronic diseases as diabetes and HIV infection. Recent advances in wilderness and battlefield medicine have reduced the risk of gangrene in injured hikers, explorers, and military personnel. Current clinical trials are evaluating the effectiveness of new antibiotics in treating diabetic foot gangrene and of hyperbaric oxygen therapy in treating gas gangrene.

SEE ALSO Diabetes; Frostbite; Necrotizing fasciitis

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# m

# Gastroesophageal Reflux Disease

# Definition

Gastroesophageal reflux disease, or GERD, is a chronic condition in which the backward flow (reflux) of stomach acid into the esophagus leads to heartburn, chest pain, and possible long-term health complications. The underlying cause is weakness in the sphincter (ring-shaped muscle) at the lower end of the esophagus where the esophagus joins the stomach.

# Description

GERD could be described as a more serious or chronic form of gastroesophageal reflux (GER), a condition that occurs when the lower esophageal sphincter (LES) opens by itself for varying periods of time or does not close properly. When the LES is open, the contents of the stomach move upward into the esophagus. The acid in the digestive juices irritates the tissues that line the esophagus, causing a burning sensation behind the breastbone or at the back of the throat. If the stomach contents are regurgitated (brought back up without trying) as far as the mouth, the person will experience a sour or unpleasant taste in the mouth.

Most people have occasional episodes of heartburn (also called acid indigestion) because of emotional stress, something they ate, or eating too large a meal. The time to be concerned about GERD is when heartburn occurs more than twice a week, is severe enough to wake the person from sleep, or is not helped by over-the-counter antacids.

GERD is not just a problem for adults; it can affect children as well. One major difference between children and adults with GERD, however, is that children are more likely to develop GERD without heartburn. Instead, their symptoms are more likely to include a dry cough, bad breath, trouble swallowing, or wheezing. In babies, symptoms of GERD may include spitting up food repeatedly, failure to gain weight, burping, and refusing food.

# Demographics

Heartburn is a very common digestive problem in the general population. According to the National Institutes of Health (NIH), more than

# Also Known As

GERD, chronic heartburn

#### Cause

Stomach acid rising into the esophagus

#### Symptoms

Recurrent heartburn, chest pain, difficulty swallowing

#### Duration

Months or years unless treated

60 million Americans experience heartburn at least once a month. Some studies indicate that more than 15 million Americans experience heartburn symptoms every day.

About 35 percent of babies born in the United States develop GERD during their first few months of life, mostly because their digestive systems are not completely developed. Most of these infants, however, will outgrow GERD by the time of their first birthday.

Among adults, GERD is most common in people over forty. It appears to affect all races and ethnic groups equally. Uncomplicated GERD is equally common in men and women. Men, however, are three times more likely than women to develop a chronic inflammation of the esophagus, and ten times more likely to



develop Barrett esophagus, a precancerous change in the cells of the tissues at the lower end of the esophagus.

Illustration of GERD, in which the contents of the stomach bubble up into the esophagus. © PHOTOTAKE INC. / ALAMY.

Some people are at increased risk of developing GERD:

- Pregnant women.
- Obese people.
- Smokers.
- People with a hiatal hernia. A hiatal hernia is a condition in which the upper part of the stomach pushes upward through a weak spot in the diaphragm. The hernia weakens the ability of the lower esophageal sphincter to keep stomach acid from flowing into the esophagus.
- People who eat large amounts of foods known to increase the amount of acid in the stomach. These include citrus fruits, chocolate, tea, coffee, alcohol, fatty and fried foods, garlic and onions, mint flavorings, spicy foods, and tomato-based foods like spaghetti sauce, chili, and pizza.
- People who take certain types of prescription medications, most commonly tranquilizers, sleeping medicines, and medications for high blood pressure.

# When to See the Doctor about Heartburn

Occasional episodes of heartburn do not necessarily mean that someone has GERD. To tell whether a visit to the doctor for further evaluation might be a good idea, the American College of Gastroenterology (ACG) suggests the following checklist:

- Does the person have one or more of the following: pain behind the breastbone moving upward from the stomach; burning sensation in the back of the throat; or a sour taste in the mouth?
- Do these symptoms usually appear after a meal?
- Does the person experience heartburn two or more times per week?
- Do antacids provide only temporary relief from the symptoms?
- Is the person still having heartburn in spite of taking prescription medication for it?
- Does the person wake up at night because of heartburn?
- Does the person have trouble swallowing food?
- Does the person notice blood in the stools? Are they regurgitating blood?
- Is the person losing weight without trying to?

If the person can answer yes to two or more of these questions, he or she should see a doctor to be tested for GERD.

# **Causes and Symptoms**

The basic cause of GERD is the inability of the LES to keep the contents of the stomach from moving backward into the lower end of the esophagus. The weakness of the lower esophageal sphincter may result from a structural disorder like hiatal hernia (a stomach abnormality); conditions that put pressure on the contents of the stomach, like pregnancy or obesity; a digestive tract that is still developing; or a disorder of the stomach that prevents it from emptying at a normal rate of speed.

Recurrent heartburn is the most common symptom of GERD, although most children and some adults with GERD do not have it. Other symptoms include:

- Belching or burping
- Regurgitating undigested food after meals
- Nausea and vomiting; vomiting blood
- Hoarseness, particularly in the morning
- Sore throat
- Coughing or wheezing
- Difficulty swallowing

# Diagnosis

There is no single laboratory test that a doctor can use to diagnose GERD. In most cases the patient's history and description of symptoms are enough to suggest the diagnosis and begin treatment with medications and lifestyle changes.

If the patient's symptoms are severe or are not helped by initial treatments, the doctor may refer the patient to a gastroenterologist, a doctor who specializes in disorders of the digestive tract.

A gastroenterologist may order one or more of the following tests:

• Barium swallow. In a barium swallow, the patient is given a chalky liquid containing barium, a chemical that coats the inside of the

digestive tract and outlines its shape on an x ray. A barium swallow can help to detect hiatal hernias, abnormal narrowing of the esophagus, or a growth in the esophagus.

- Endoscopy. Endoscopy is a technique that allows a gastroenterologist to look directly into the esophagus and stomach by inserting a long flexible tube (endoscope) attached to a light source and video camera down the patient's throat. Endoscopy allows the doctor to take tissue samples to check for a Barrett esophagus or cancer of the esophagus as well as to look at the structure of the esophagus and LES.
- Acid probe test. This test measures the acidity of the patient's stomach contents over a 24-hour period and the length of time that the lower esophagus is exposed to stomach acid. A probe is inserted through the patient's nose via a long, flexible catheter to a point just above the LES. The other end of the catheter is attached to a small computer that the patient wears around the waist during the test. The computer measures the length of time and frequency of acid reflux into the lower esophagus.
- Tests to measure the speed of stomach emptying. These tests are usually performed only when the doctor thinks that delayed emptying of the stomach is a factor in the patient's GERD.

# Treatment

Most patients with GERD can be successfully treated by a combination of medications and lifestyle changes.

There are several types of medications that doctors may prescribe for GERD.

- Over-the-counter antacids, such as Alka-Seltzer, Maalox, Mylanta, Rolaids, and Riopan. Antacids can be purchased in any pharmacy in either tablet or liquid form and work well to control mild cases of GERD. They should be taken after each meal and at bedtime.
- Foaming agents. Gaviscon is the best-known of this type of medication. They work by coating the stomach contents with foam, which prevents reflux.
- H2 blockers. These are drugs like Tagamet, Zantac, and Pepcid; they work by decreasing the production of stomach acid. They are available in both over-the-counter and prescription strength.

- Proton pump inhibitors (PPIs). These drugs also work by decreasing stomach acid and are generally more effective than the H2 blockers. Most are available by prescription. PPIs include drugs like Prilosec, Protonix, Prevacid, and Nexium.
- Prokinetics. These are drugs that work by speeding up the rate of stomach emptying. Reglan and Urecholine are examples of drugs in this group.

People who are not helped by medications may need surgery to treat GERD. The operation that is usually done is called fundoplication. In this procedure, the surgeon wraps the upper part of the stomach around the lower end of the esophagus to strengthen the LES, prevent acid reflux, and repair a hiatal hernia. The operation is safe and can be done in infants as well as adults.

# Prognosis

Most people diagnosed with GERD do very well with medications and lifestyle changes. Of those who require surgical treatment, 92 percent have no more symptoms of GERD.

Complications from GERD, such as narrowing of the esophagus or Barrett esophagus, develop in about 20 percent of patients. These patients should be treated with surgery as soon as their complication is diagnosed.

# Prevention

Lifestyle changes are the most effective form of prevention for GERD. The NIH recommends:

- Not smoking. Smoking increases the production of stomach acid.
- Keeping one's weight within the recommended guidelines for one's age, sex, and height.
- Avoiding foods and beverages that trigger acid indigestion.
- Eating small frequent meals rather than three large ones.
- Avoiding lying down for three hours after eating.
- Raising the head of the bed by 6–8 inches (15–20 centimeters). This should be done by using wooden blocks or foam wedges; just using extra pillows will not be effective.
- Wear clothing with loose waistlines. Tight belts or waistbands put pressure on the abdomen.

# WORDS TO KNOW

- **Diaphragm:** The flat sheet of muscle that separates the abdomen from the chest cavity.
- **Esophagus:** The muscular tube that carries food downward from the lower throat to the stomach.
- **Fundoplication:** A surgical procedure in which the upper part of the stomach is wrapped around the lower end of the esophagus to prevent stomach acid from rising into the esophagus.
- **Gastroenterologist:** A doctor who specializes in diagnosing and treating disorders of the digestive system.

- **Hiatal hernia:** A condition in which the upper part of the stomach bulges upward into the chest cavity through a weak spot in the diaphragm.
- **Reflux:** The medical term for the backward flow of stomach acid from the stomach into the esophagus.
- **Regurgitation:** Throwing up; effortless flow of undigested stomach contents back up the eso-phagus into the mouth.
- **Sphincter:** A ring-shaped muscle that can contract to close off a body opening.

# **The Future**

A surgical treatment for GERD that was approved by the Food and Drug Administration (FDA) in 2000 is the use of an endoscope to insert stitches into the LES to strengthen the muscle or to use radiofrequency energy to form scar tissue on the LES to tighten it. However, the long-term effectiveness of this type of surgery compared to fundoplication is not known.

SEE ALSO Alcoholism; Obesity; Smoking; Stress; Ulcers

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# Genetic Disorders

Genetic disorders are a group of lifelong diseases caused by abnormalities in genes or chromosomes that are present at the time that a baby is conceived. Genetic disorders may involve extra chromosomes (Down syndrome, Edwards syndrome, Patau syndrome, triple X syndrome, Klinefelter syndrome), a missing chromosome (Turner syndrome), or mutations in the parents' sperm or egg (fragile X syndrome, Huntington syndrome).

About 4,000 genetic disorders are known to exist. Most of these are very rare, affecting only one person in several thousand or several million. Cystic fibrosis, the most common genetic disorder in the United States, affects one child in every 3,900. However, about one American in every twenty carries the gene for the disease.

Genetic disorders vary in their inheritance patterns. In some disorders, like Huntington disease or Marfan syndrome, the child needs only one copy of the mutated gene to develop the disorder. In others, such as cystic fibrosis or sickle cell anemia, the child must inherit the defective gene from both parents in order to have the disease. Some genetic disorders are caused by defective genes on the X or Y sex chromosomes.

**SEE ALSO** Achondroplasia; Cystic fibrosis; Down syndrome; Edwards syndrome; Fragile X syndrome; Huntington disease; Hutchinson-Gilford syndrome; Klinefelter syndrome; Marfan syndrome; Patau syndrome; Phenylketonuria; Sickle cell anemia; Thalassemia; Triple X syndrome; Turner syndrome; Xeroderma pigmentosum



# Genital Herpes

# Definition

Genital herpes is a sexually transmitted disease (STD) caused by a virus group that may also cause cold sores in the mouth. It is contagious and can be spread by carriers—persons who are infected with the virus but have no symptoms of the disease. As many as 80–90 percent of people infected by the herpes virus are asymptomatic carriers.

# Description

Genital herpes is a sexually transmitted viral infection whose most noticeable symptom is blisters or open sores on the skin of the genitals. It is caused by the herpes simplex virus (HSV), which has two forms, known as HSV-1 and HSV-2. HSV-1 is familiar to many people as the virus that causes cold sores around the outside of the mouth; it is responsible for some cases of genital herpes as well. HSV-2 causes most cases of genital herpes; it is spread by mouth-to-genital or genital-to-genital contact. HSV is shed from visible sores, blisters, or a rash during outbreaks of genital herpes, but can also be shed from the affected area between outbreaks of symptoms.

HSV can be transmitted even when the infected person has no symptoms; in other words, a sexual partner without obvious genital sores or blisters can still transmit the illness. People without noticeable symptoms may actually contribute more to the spread of genital herpes than those with active sores. Also Known As Herpes

#### Cause

Virus

#### Symptoms

Pain, itching, blisters or open sores on genitals, fever, headache

#### Duration

May cause intermittent episodes for years



Image of the herpes simplex virus, which causes genital herpes. JAMES CAVALLINI / PHOTO RESEARCHERS, INC.

More than some other sexually transmitted diseases (STDs), herpes can be a source of severe emotional distress to people infected with it. These feelings—particularly fear of rejection—are largely related to the fact that unlike such STDs as chlamydia and gonorrhea, genital herpes presently has no permanent cure. This fact means that anyone who is in a sexual relationship with an infected person runs some risk of being infected themselves even when the partner is being treated for HSV infection. Some people do choose to end relationships with infected partners rather than run this risk.

# **Demographics**

Genital herpes is an extremely common infection among sexually active adults around the world. The rate has been increasing in most developed countries in recent years although it has recently decreased in the United States. According to the Centers for Disease Control and Prevention (CDC), there are between 45

and 50 million people infected in the United States, with an estimated one million new infections every year. About one in every five sexually active adolescents and adults are infected with HSV. Women are about twice as likely to be infected with herpes by having sex with men than men are to get the infection from a woman.

Some groups of people are at greater risk than others of being infected with genital herpes. High-risk groups in the United States include:

- Young adults of either sex.
- People who have unprotected sex or do not use barrier methods of birth control (condoms or diaphragms).
- Homosexual men, particularly those who are HIV-positive or abuse drugs or alcohol.
- People living in large cities.
- People with low income or little education.
- People who use cocaine.
- People who have a large number of sexual partners.

# **Causes and Symptoms**

Genital herpes is caused by herpes simplex virus (HSV). It may be caused by either HSV-1, which also causes cold sores around the mouth, or primarily by HSV-2. People who have never had cold sores may have a prodrome, or period when they have some warning symptoms of the disease. The prodrome of genital herpes may be marked by flu-like symptoms, including loss of appetite, fever, and a general unwell feeling. Some people feel pain, itching, burning, or a tingling sensation in the areas where the blisters of herpes erupt several days later.

Although genital herpes is usually transmitted by mouth-to-skin contact or sexual activity, it can also be transmitted by a pregnant woman to her baby during childbirth. If a woman has active genital herpes at the end of her term, the baby is usually delivered by cesarean section. Fortunately, infection of a baby from a woman with herpes infection is rare. There have also been cases of HSV transmission through skin-to-skin contact in such sports as wrestling or rugby.

# After the prodromal period, infected people of either sex typically notice what are often repeated eruptions of small aching blisters (between onequarter and one-half in size) filled with clear yellowish fluid on the genitals, around the rectum, or covering nearby areas of skin. The blisters typically occur in clusters or crops. The blisters then break, leaving shallow open sores that are very painful. These sores eventually crust over and slowly heal over a period of two to four weeks. The patient's lymph glands may also become sore and swollen. Both men and women may experience pain when urinating, and women may notice a vaginal discharge.

After a person is infected with HSV, the virus hides within nerve cells, making it difficult for the immune system to find and destroy it. The virus remains in the body, so that symptoms can reappear at any time. Recurrences of genital herpes can be triggered by a number of factors. The most common triggers, however, are emotional stress; menstruation (in women); inadequate sleep and fatigue; another illness; a surgical procedure; and irritation of the skin from clothing or athletic gear.

# Self-Care for Genital Herpes

People diagnosed with genital herpes can ease some of the symptoms of an outbreak by the following self-care measures:

- Avoiding sexual contact with others until the sores have healed.
- Keeping the sores cool, clean, and dry.
- Washing the hands after any contact with the sores.
- Practicing safe sex.
- Wearing loose and comfortable cotton clothing; tight garments and synthetic fabrics can irritate the sores.
- Avoiding the use of heavily perfumed or antibacterial soaps, bath gels, or feminine hygiene products.
- Taking aspirin, Motrin, Advil, or Tylenol for pain or fever.

# Diagnosis

The diagnosis of genital herpes is often delayed, particularly if the person has had only mild symptoms that can be mistaken for those of another disease. Herpes can be detected by a blood test or by a laboratory culture made from fluid or a tissue scraping from a blister or sore. A tissue culture generally takes several days to give results.

Many people who do have symptoms of genital herpes are also infected with HIV, gonorrhea, syphilis, or other STDs. It is now common for doctors to test patients for these other diseases to make sure which disease is causing the patient's symptoms and that all the STDs that are present are being treated.

# Treatment

Treatment for genital herpes consists of one of several antiviral medications taken by mouth to lower the risk of relapses, to lower the risk of spreading herpes, and to heal sores and blisters more rapidly. The most common medications prescribed for herpes are Valtrex, Famvir, and Zovirax. These drugs do not, however, cure the disease. They are usually prescribed for a period of seven to ten days for the first outbreak and five days for recurrences; however, patients who have frequent outbreaks may be able to control them only by taking an antiviral drug every day.

# Prognosis

There is no cure for genital herpes. Although the majority of infected persons may never have noticeable symptoms, those who do may have recurrent outbreaks as long as forty years after the initial infection. Eighty-five percent of people with symptomatic genital herpes have recurrences sometimes as many as six to ten each year. Recurrent herpes infections in men are generally milder and shorter in duration than those in women.

# Prevention

Genital herpes can be prevented in several ways:

- Abstaining from sexual intercourse or limiting intercourse to one partner who is free of sexually transmitted diseases.
- Not having sex with high-risk partners.
- Infected persons should not have sex at all when they have blisters or sores anywhere on the body or in the mouth. Even the use of a

# WORDS TO KNOW

**Carrier:** A person who is infected with a disease and can spread it to others but who has no symptoms of the disease.

**Prodrome:** A period before the acute phase of a disease when the patient has some characteristic warning symptoms.

condom does not prevent the spread of herpes when someone has genital sores or blisters because not all sores will be covered by the condom.

- Using latex condoms every time during sex when sores or blisters are not present.
- Avoiding the use of drugs (particularly cocaine) and alcohol, which can impair good judgment.
- Having sexual partners tested or treated for herpes before having sexual relations.

# **The Future**

The number of new herpes infections is likely to continue to rise worldwide, if only because the majority of infected persons are carriers rather than having obvious symptoms of the disease. One possible treatment that might reduce the risk of symptomatic herpes in women is a vaccine against HSV-2 called Herpevac, which was being tested by the National Institutes of Health (NIH) as of 2008. Results so far indicate that the vaccine would not help everyone; it does not prevent genital herpes in men, and it works only in women who have never been exposed to HSV-1. The partial success of Herpevac does, however, encourage researchers to continue working on vaccines against genital herpes.

SEE ALSO AIDS; Chlamydia; Cold sore; Gonorrhea; Syphilis

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# German Measles

# See Rubella.

# <mark>m</mark> Gigantism

# Definition

Gigantism is a term for abnormal growth in height in children caused by too much growth hormone. An oversupply of growth hormone in adults who have completed their skeletal growth leads to a condition called acromegaly.

Gigantism in children and adolescents is based on a comparison of the child's height and weight to those of others of his or her age, sex, and level of sexual maturity; there is no absolute minimum height or weight that defines gigantism.

# Description

Gigantism in children can begin at any age before the growth plates in the long bones of the body close, usually by the early twenties. It is often quite dramatic because parents typically notice their children's growth and because the growth plates in the arms and legs respond quickly to growth hormone. The tallest man ever recorded, Robert Wadlow (1918–1940) of Alton, Illinois, began growing abnormally rapidly at the age of four. By age 10 he was 6 feet 6 inches tall (2 meters); by age 16 he was 7 feet 11 inches tall (2.4 meters); at the time of his death at age 22 he was 8 feet 11 inches (2.7 meters) tall and reportedly still growing.

#### Also Known As

Pituitary gigantism; acromegaly (in adults)

#### Cause

Overproduction of growth hormone

#### Symptoms

Unusually rapid growth, delayed puberty, shortened lifespan if untreated

#### Duration

Years

#### Gigantism



Sandy Allen with her little brother, of normal stature. Sandy was the second tallest woman in the world when she died in August 2008. BETTINA CIRONE / PHOTO RESEARCHERS, INC.

Acromegaly in adults, by contrast, is much slower in onset, is not recognized as quickly, and may continue for some years before it is diagnosed. Most adults with acromegaly begin to develop symptoms in their twenties but diagnosis is delayed on average for about nine years, with some patients not diagnosed for fifteen years. The average age at diagnosis is forty years.

## Demographics

Both gigantism and acromegaly are rare disorders, although gigantism is much rarer than acromegaly. There are only about a hundred known cases of gigantism reported in medical journals, whereas acromegaly affects between forty and seventy adults per 1 million population.

As far as is known, males and females are equally affected by gigantism and acromegaly, as are members of all races and ethnic groups.

# **Causes and Symptoms**

Gigantism in children is caused by too much growth hormone produced by the pituitary gland, which is a peanut-shaped gland that lies at the base of the brain behind the nose. There are several reasons why this gland may start overproducing. The most common cause is a benign (noncancerous) tumor in the pituitary gland that leads to the secretion of too much hormone. The second reason, which is much less common, is one of several

# André the Giant (1946-1993)

André the Giant was the stage name of a professional wrestler and movie actor named André Roussimoff. He was born in Coulommiers, France, to parents of Bulgarian and Polish descent. He quit school after eighth grade and worked as a farm laborer and factory worker for several years. He was then discovered by a wrestling promoter and moved to Paris to train as a professional wrestler. By the time he was 21, he had become a European wrestling star. He traveled to Japan for a wrestling match in 1969; at that time he was diagnosed by a Japanese doctor as having acromegaly.

In 1973 André began his wrestling career in North America and took the stage name of "André the Giant." He became an actor as a sideline to wrestling. He starred in several films and television series in the 1970s and 1980s, including the role of Fezzik in *The Princess Bride*, which was his favorite part. By 1987, however, André's weight had reached 560 pounds, and his bones and joints were causing him great pain. Although he continued to wrestle professionally until 1991, his health gradually failed. He died in his sleep in



Andre the Giant in 1983. MICHAEL ABRAMSON/LIAISON.

January 1993 of congestive heart failure after returning to Paris to attend his father's funeral. His death was attributed to the acromegaly.

rare medical conditions that also affect the pituitary gland. The third cause, which is very rare, is that the child has an oversupply of a protein that increases the activity of a normal supply of growth hormone.

Acromegaly in adults is caused by a tumor in the pituitary gland leading to an oversupply of growth hormone in 95 percent of cases; however, there are also cases of acromegaly caused by tumors in the pancreas, adrenal glands, or lungs. These tumors may actually secrete growth hormone, or they may secrete another hormone that stimulates the pituitary gland to produce more growth hormone.

The symptoms of gigantism and acromegaly include the following:

- Rapid increase in height (in children)
- Increase in weight leading to mild or moderate obesity

- Headaches and vision problems caused by the pressure of a pituitary tumor on the optic nerve and other parts of the brain
- Exaggeratedly large hands and feet with unusually thick fingers and toes
- Coarse facial features with an enlarged jaw and rounded swelling of the forehead
- Thick, oily skin with heavy sweating and body odor
- · Deepened or husky voice, tendency to snore heavily at night
- · Joint problems leading eventually to osteoarthritis
- Increased risk of other benign tumors and diabetes
- Delayed puberty in children; sexual problems in adults
- Enlarged liver, heart, spleen, and kidneys
- Overgrowth of soft tissue

# Diagnosis

The diagnosis of gigantism or acromegaly is made on the basis of the patient's history and a series of laboratory tests and imaging studies. The most common is a blood test taken after the patient has fasted overnight, which evaluates the level of growth hormone and another hormone secreted by the liver in response to growth hormone. If the levels of both hormones are high, the test indicates gigantism or acromegaly.

A second blood test that may be performed to confirm the diagnosis is called a growth hormone suppression test. The patient has two blood samples taken, one before and one after drinking a sugar solution. In a normal person, the level of growth hormone will drop after drinking the sugar solution. If the level stays high, it confirms a diagnosis of gigantism or acromegaly.

The doctor will also usually order a CT scan or MRI in order to determine whether there is a tumor in the patient's pituitary, or whether a tumor elsewhere in the body may be responsible for the high levels of growth hormone. In a few cases the patient may also need a chest x-ray to help locate the tumor.

# Treatment

The most common treatment for gigantism and acromegaly is surgical removal of the pituitary tumor. This is a complicated procedure requiring a highly experienced surgeon. In some cases the surgeon cannot remove the entire tumor. The patient is then treated with synthetic hormones that block the production of growth hormone, or with a newer drug that blocks the effect of growth hormone on body tissues. The patient must inject these drugs at home on a daily basis.

Patients whose tumor cannot be completely removed by surgery may be given radiation therapy. This type of treatment is considered a last resort, however, as it usually upsets the balance of other hormones in the body and can cause learning difficulties, obesity, and personality changes in children. In addition, radiation therapy may take from eighteen months to several years to stop the overproduction of growth hormone and requires frequent follow-up treatments for the rest of the patient's life.

## Prognosis

The prognosis for patients with gigantism or acromegaly is usually good to excellent following surgery; smaller tumors can be removed successfully about 85 percent of the time and larger tumors between 50 and 65 percent of the time. About 13 percent of children with pituitary tumors have a recurrence of the tumor after surgery, however.

With regard to medications, some patients do not respond to the drugs that are currently used. Radiation therapy can cause complications by suppressing the secretion of other body hormones as well as fighting the cells in a pituitary tumor. About 60 percent of patients who are treated with radiation therapy for acromegaly must take hormone replacements for the rest of their lives.

Although gigantism and acromegaly are not fatal by themselves, they can cut short a patient's life span if they are not treated because of the extra strain put on the lungs and heart by the patient's body size. Patients with untreated acromegaly usually develop high blood pressure (hypertension) and heart problems. They are also at increased risk of diabetes, breast or colon cancer, osteoarthritis, vision loss, carpal tunnel syndrome, spinal cord compression, and uterine polyps (in women).

## Prevention

There is no known way to prevent gigantism or acromegaly because the factors that lead to pituitary tumors are not well understood. It is known that gigantism and acromegaly are not hereditary, because patients with these conditions typically have parents and siblings of normal height and weight.

# WORDS TO KNOW

- Acromegaly: A condition in which a person's body produces too much growth hormone in adult life.
- **Gigantism:** Excessive production of growth hormone in children who are still growing.
- **Growth plate:** A cartilage plate in the long bones of children where the lengthening of bone takes place.

**Pituitary gland:** A pea-sized gland located at the base of the brain behind the nose that secretes growth hormone and other hormones that affect sexual development and the body's response to stress.

# The Future

Gigantism and acromegaly are both rare disorders and not likely to increase in frequency over the next few decades. It may be that improved surgical techniques or the development of new medications will make it easier to treat these conditions without the side effects that many patients presently have to accept. Two newer drugs were at the clinical trial stage as of early 2008.

**SEE ALSO** Childhood obesity; Diabetes; Heart failure; Hypertension; Osteoarthritis

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# <mark>m</mark> Glaucoma

# Definition

Glaucoma is not a single eye disorder but a group of disorders that affect the eye. The optic nerve is made up of 1 million nerve fibers. It carries visual images from the retina, the layer of light-sensitive tissue at the back of the eyeball, to the brain. Untreated glaucoma may lead to vision loss or even complete blindness. In most cases of glaucoma, the optic nerve is damaged by increased fluid pressure inside the eyeball. However, reduced blood supply to the optic nerve, caused by the increased pressure, can also be a factor.

Glaucoma is often categorized as either primary or secondary. Primary glaucoma refers to glaucoma that is not triggered by an injury or other medical condition. It accounts for about 90 percent of cases. There are several major types of primary glaucoma:

- Open-angle glaucoma. This term refers to the angle in the front portion of the eye where the cornea meets the iris. In normal circumstances, fluid flows in and out of the front of the eye through a meshwork of tissue at the angle that acts like a drain. In openangle glaucoma, the angle remains open and fluid continues to pass through the meshwork, but not quickly enough. As a result, fluid builds up inside the eye, and the fluid pressure may rise high enough to damage the optic nerve. Open-angle glaucoma is the most common type of primary glaucoma.
- Closed-angle glaucoma. Closed-angle glaucoma is a disorder in which the angle between the iris and the cornea is blocked, usually because the iris becomes swollen from pressure and moves forward to touch the meshwork directly, thus preventing fluid from draining out of the eye. Closed-angle glaucoma can develop either gradually or suddenly. Acute closed-angle glaucoma is a medical emergency.
- Congenital glaucoma. This is a type of glaucoma that is present at birth and is usually noticeable within the first year of life. It is more common in boys than in girls. In congenital glaucoma, there is a defect in the structure of the baby's eye that slows down the normal drainage of fluid.

#### Cause

Increased pressure inside the eye

#### Symptoms

Blurred vision, eye pain, seeing halos around lights, tunnel vision

#### Duration

May develop over years or within hours, depending on type

*Eye showing acute glaucoma.* © MEDICAL-ON-LINE / ALAMY.



- Normal-pressure glaucoma. Also called normal-tension glaucoma, this is a type of primary glaucoma in which the optic nerve is damaged even though the fluid pressure within the eye is within normal range. Doctors do not yet understand the causes of this type of glaucoma, although one theory suggests that the optic nerve is damaged by normal fluid pressure because its blood supply has been reduced.
- Pigmentary glaucoma. This is a type of glaucoma that develops when pigment granules from the iris flake off and block the drainage meshwork in the angle between the iris and cornea.

About 10 percent of cases of glaucoma are secondary, meaning that they develop as a result of injury to the eye or another disease, most commonly diabetes, leukemia, or sickle cell anemia. Secondary glaucoma may result from a blow to the eye, a tumor, cataract surgery, or the use of corticosteroid medications.

# Description

People's experience of glaucoma varies considerably depending on which type they have and whether it is chronic (developing slowly) or acute (sudden onset). In addition, glaucoma can affect both eyes or only one.

# When to See the Doctor

Some types of glaucoma develop gradually and can usually be detected during routine periodic eye examinations. Acute closed-angle glaucoma, however, is a medical emergency. Anyone with any of the following symptoms should see an eye doctor *at once*:

- Blurred vision
- Severe pain in the affected eye or eyes
- Redness in the eye
- Sudden violent headache
- Abdominal cramping
- Seeing rainbow-like halos around lights

Emergency treatment is needed because this type of glaucoma can cause permanent blindness in as little as one or two days.

Children with congenital glaucoma are usually diagnosed within the first few months after birth because their eyes look cloudy, are unusually sensitive to light, and secrete large amounts of tears.

Chronic open-angle glaucoma, the single most common type, develops over a period of years and is related to the aging of the drainage meshwork in the angle between the iris and cornea. In the early stages, patients with this type of glaucoma may have no symptoms at all. Gradually, however, they find it more difficult to see objects to the side or on the edges of their visual field. They may also develop tunnel vision, in which they can see only objects straight in front of them.

Closed-angle glaucoma can come on suddenly, often in dim light, with eye pain, reddening of the eye, a sudden severe headache, and nausea and vomiting. The person may also see colored or rainbow-like halos around lights.

# **Demographics**

Glaucoma is largely an eye disorder of adults. Congenital glaucoma is rare and childhood glaucoma is also unusual. Glaucoma in middle-aged adults, however, is a common eye disorder and the second leading cause of blindness in the United States. According to Prevent Blindness America, more than 2.2 million Americans over age forty have open-angle glaucoma, but only half of them know that they have it. In North America and Europe, glaucoma affects one person in every two hundred aged fifty or younger, but one in ten over the age of eighty.

Some people are at increased risk of glaucoma:

- Those with increased fluid pressure in the eye, sometimes called ocular hypertension. A high level of fluid pressure inside the eye is the greatest single risk factor for glaucoma. However, as noted earlier, some people develop the disorder even though their eye fluid pressure is normal.
- Age. The risk of glaucoma increases over age sixty for most Americans; it rises over age forty for African Americans.

- Race and ethnicity. African Americans have six to eight times the risk of glaucoma as Caucasians. Mexican Americans over the age of sixty are also at increased risk. Asian Americans and Alaskan Inuit have a higher than average risk of acute closed-angle glaucoma, and Japanese Americans have an increased risk of normal-pressure glaucoma. The reasons for these differences are not yet understood.
- Family history of glaucoma.
- Certain diseases or conditions, including high blood pressure, diabetes, heart disease, hypothyroidism, and sickle cell anemia.
- Myopia (nearsightedness).
- A history of injury to the eye, inflammation of the eye, tumors in the eye, or cataract surgery.
- Long-term use of corticosteroid medications.

# **Causes and Symptoms**

The most important cause of glaucoma is increased fluid pressure within the eye resulting from overly slow drainage of fluid through the meshwork in the angle between the iris and cornea or complete blockage of the angle. This buildup of fluid pressure damages the optic nerve. In some cases inadequate blood supply to the optic nerve is also a factor.

There is some evidence that genetic factors are also involved in glaucoma because the disorder is known to run in some families. Several genetic mutations have been linked to primary open-angle glaucoma, but no single gene has been shown to cause the disorder.

The symptoms of the various types of glaucoma were described earlier.

# Diagnosis

The diagnosis of chronic primary glaucoma is usually made during a routine eye examination by an ophthalmologist, who is a doctor specializing in the diagnosis and treatment of eye disorders. A complete eye examination, depending on the individual patient's history and risk factors, will include most or all of the following tests:

• Dilation of the eye. The doctor will dilate the pupil of the eye by placing drops in the eyes that keep the iris from narrowing the pupil when the doctor shines a bright light directly into the eye.

The doctor will then be able to see directly to the back of the eye to check for damage to the optic nerve.

- Tonometry. This is a test for measuring the level of fluid pressure inside the eye. It can be performed either by resting an instrument briefly on the surface of an anesthetized eye, or by blowing a puff of air onto the surface of the eye while the patient's chin is held steady. The fluid pressure is estimated by measuring the response of the eye to the puff of air.
- Tests of peripheral vision. Since open-angle glaucoma often starts with gradual loss of side vision, doctors may measure whether such loss has occurred by asking patients to look at a set of blinking lights and indicate when they can see the lights. The patients' answers allow doctors to map how much, if any, of the patients' visual field has been lost.
- Tests to measure the thickness of the cornea. This test is done to rule out the possibility that a cornea that is either unusually thick or unusually thin is affecting the measurement of the fluid pressure inside the eye.
- Gonioscopy. This is a test than uses a gonioscope, an instrument with a mirror as well as a light source. It allows the doctor to tell whether the angle between the iris and the cornea is open or closed.

A patient with acute closed-angle glaucoma will usually be treated by an ophthalmologist in a hospital emergency department. Emergency treatment consists of medications to quickly reduce the fluid pressure inside the eye and laser surgery to cut a drainage hole in the iris.

# Treatment

Glaucoma can be treated with medications, surgery, or both. The medications used include special eye drops or oral medications. In some cases the patient may be asked to use more than one type of eye drop. There are several different types of drugs that may be prescribed; some work to lower the fluid pressure inside the eye by decreasing the amount of fluid produced. Other drugs work by increasing the outflow of fluid. All of these medications have side effects, however, and must be used exactly according to the doctor's instructions.

Surgery is usually used to treat congenital glaucoma and acute closedangle glaucoma. The procedure used to treat closed-angle glaucoma is called an iridotomy. The surgeon uses a laser to cut a hole in the iris to relieve the increased pressure inside the eye.

Surgery can also be performed to treat open-angle glaucoma. There are two major types of laser surgery that can be done. One technique involves using the laser to open clogged portions of the drainage network. Another approach is called filtering surgery. The surgeon uses the laser to cut a small hole in the sclera (white part of the eyeball) and remove a small portion of the clogged meshwork. The extra fluid can then leave the eye through the hole in the sclera.

Children with glaucoma and adults with secondary glaucoma can be treated with drainage implant surgery. The implant is a small silicone tube that the doctor inserts inside the eye to help drain the excess fluid.

# Prognosis

The prognosis of glaucoma depends on its type and the stage at which it is diagnosed. Vision that has been lost to any type of glaucoma cannot be restored. Without treatment, acute glaucoma results in permanent vision loss within days. Untreated chronic glaucoma can progress to blindness within several years.

# Prevention

The best prevention for glaucoma is regular eye exams. Anyone over age eighteen should be screened periodically for glaucoma. The schedule of exe examinations recommended by the National Eye Institute (NEI) is as follows:

- Eighteen to sixty years of age: Every two years.
- Over sixty: Every year.
- One or more risk factors other than age: Every year before and after age sixty.

Other preventive steps that people can take include getting treatment (special eye drops) if diagnosed with high fluid pressure inside the eye; controlling one's weight; and getting treatment for high blood pressure.

# The Future

Glaucoma is likely to become more common in the general population because of the growing number of people over age fifty. Present research is focused on identifying more genes associated with glaucoma; developing better treatments for high fluid pressure in the eye before it leads

# WORDS TO KNOW

- **Cornea:** The transparent front part of the eye where light enters the eye.
- **Iris:** The circular colored structure at the front of the eyeball that controls the amount of light entering the eye by changing the size of the pupil.
- **Ophthalmologist:** A doctor who specializes in diagnosing and treating eye disorders and can perform eye surgery.
- **Pupil:** The circular opening in the center of the iris.
- **Retina:** The light-sensitive layer of tissue at the back of the eyeball.
- **Sclera:** The opaque white portion of the eyeball.

to glaucoma; and comparing the effectiveness of the various surgical treatments for glaucoma.

**SEE ALSO** Cataracts; Diabetes; Hypertension; Leukemia; Myopia; Sickle cell anemia

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# Gonorrhea

# Definition

Gonorrhea is an infectious disease caused by bacteria called *Neisseria gonorrhoeae*, or gonococcus. The bacteria are transmitted by sexual intercourse or, now rarely in the Western world, from an infected mother to her baby during childbirth.

# Description

Gonorrhea is a sexually transmitted disease (STD) that causes inflammation and a discharge from the urinary tract and the genitals, although it can also cause infections of the throat, eyes, and rectum. Babies can be infected before or during childbirth. In addition to the immediate symptoms of infection, gonorrhea can cause dangerous long-term complications if it is not diagnosed and treated. It can cause blindness in newborns, infertility in adult women, inflammation of the prostate gland in men, and joint infection. Other rare long-term complications include inflammation of the tissues covering the brain and spinal cord or infection of the heart valves.

# Demographics

Gonorrhea is the most common infectious disease that doctors in the United States are required to report to public health officials. It is estimated that there are 600,000 new infections in the United States each year; there are about 240 cases reported per 100,000 people. The rates are considerably higher among African Americans than among other racial groups, particularly in inner-city areas and in the southeastern United States. The lowest rates are among Asian Americans and Pacific Islanders.

Age is a major factor in the spread of gonorrhea. Three out of four cases of the disease are found in people younger than thirty. The highest rates of gonorrhea are found in women between fifteen and nineteen years of age and in men between the ages of twenty and twenty-four.

Other risk factors for getting gonorrhea are a large number of sexual partners and having other sexually transmitted diseases (STDs). Gonorrhea in children almost always points to sexual abuse. Also Known As Gonococcal infection, the clap

Cause Infection by a bacterium

#### Symptoms

Genital discharge, painful or frequent urination, pain or bleeding during intercourse

#### Duration

Symptoms begin two to ten days after exposure



Newborn with eyes infected with gonorrhea from the mother, who had the venereal disease when she gave birth. DR. M.A. ANSARY/PHOTO RESEARCHERS, INC.

# **Causes and Symptoms**

The gonorrhea bacterium is spread through contact with infected semen or vaginal fluids during unprotected sexual intercourse or during childbirth. People can become carriers of the disease without developing its characteristic symptoms. Men have a 20 percent chance of getting infected by a single sex act with an infected woman, while women have a 50 to 70 percent chance of getting gonorrhea from an infected man during a single sex act. Men who have sex with men run a high risk of getting gonorrhea during anal intercourse, particularly if either partner has HIV infection.

The bacterium enters the body in the semen or vaginal fluids of an infected partner. Once inside the vagina, mouth, or rectum, it attaches itself to the moist tissues that line those parts of the body. There it multiplies and spreads below the uppermost layer of tissue. The body responds by sending white blood cells to attack the invaders and by shedding the infected tissue. The result is inflammation and the production of a whitish or yellowish discharge. Most people develop symptoms between two and ten days after unprotected sex.

Men are more likely to develop noticeable symptoms than women. In men, the most common symptoms of gonorrhea are a sticky discharge of pus from the penis and pain when urinating. In women the most common symptoms are a vaginal discharge, pain during intercourse, bleeding after intercourse, or difficulty urinating.

Between 30 and 60 percent of infected women may not have any noticeable symptoms of infection and may fail to get tested for gonorrhea. They are at high risk of a complication of the disease called pelvic inflammatory disease or PID. In PID, the bacterium travels upward from the vagina through the uterus and the other reproductive organs. It produces inflammation that causes scarring of the tubes that carry fertilized eggs to the uterus. In addition to causing cramps, painful menstrual periods, and fever, PID can leave a woman permanently unable to have children. It can also cause a complication called an ectopic pregnancy, in which the fertilized egg starts growing inside the tubes leading to the uterus or in the abdomen rather than in the uterus itself.

Another complication of untreated gonorrhea is called disseminated gonococcal infection or DGI. Disseminated is a medical word that refers to the spread of an infection throughout the body. About 1–2 percent of people infected with gonorrhea develop DGI. The gonorrhea bacteria get into the bloodstream and are carried to other parts of the body. It can cause arthritis if it infects a joint. It can also infect the valves in the heart or the protective tissues that cover the brain and spinal cord.

# Diagnosis

Gonorrhea is usually diagnosed by taking a sample of the discharge from the patient's genitals or rectum, or a throat swab, and sending the sample to a laboratory for analysis. In addition, the doctor will take a careful history of the patient's sexual contacts in order to determine whether they may have given the disease to others, and whether they may have gotten another sexually transmitted disease as well as gonorrhea. Doctors are required by law to report cases of gonorrhea to the Centers for Disease Control and Prevention (CDC) and to local public health departments.

# Treatment

The treatment of gonorrhea has become complicated in recent years because the bacteria have developed resistance to many of the antibiotics that were formerly effective against it. In April 2007 the Centers for Disease Control and From the late Middle Ages to the early twentieth century, gonorrhea was often treated by giving the patient mercury, either by mouth, by rubbing it on the skin, or by injecting it into the opening of a man's urethra. One way of treating patients with mercury that seems strange nowadays was called fumigation. The infected person was put inside a large wooden box with only their head sticking out. A small quantity of mercury was poured into the box, which was then heated. The heat caused the mercury to vaporize and enter the body when the patient breathed the vapors.

Mercury, however, is known to be toxic, causing teeth to fall out as well as causing chest pains, insomnia, muscle cramps, hallucinations, sleep disorders, paralysis, and depression. It was a common treatment for gonorrhea in spite of its dangers. The Lewis and Clark expedition (1804–1806) carried a supply of mercury for treating members of the group who had contracted gonorrhea from infected Native American women. The poet John Keats (1795– 1821) died of tuberculosis, but some medical historians think that he weakened his lungs four years before his death by dosing himself with mercury to cure himself of gonorrhea.

As late as World War I (1914–1918) the medical department of the U.S. Army used a mercury compound to treat soldiers diagnosed with gonorrhea. Soldiers who lacked "moral stamina and self-control," as the War Department put it, could ask for a tube of ointment containing mercury and silver for use after a night on the town. It was only with the mass production of penicillin in the 1940s that doctors had a treatment for gonorrhea that was safe as well as effective. Prevention (CDC) added gonorrhea to the list of so-called superbugs that is, diseases that are resistant to many antibiotics in common use. The standard treatment for uncomplicated gonorrhea is a single dose of an antibiotic, most commonly ceftriaxone (by injection) or cefixime (by mouth).

People with symptoms of either DGI or PID may need to be treated in a hospital, particularly if the heart or central nervous system is affected.

# Prognosis

The prognosis for recovering from uncomplicated gonorrhea is very good provided that treatment is given promptly. Women have a 15 percent risk of being unable to have children after one episode of PID; if they have another attack, the risk rises to 50 percent. Most people can be successfully treated for DGI; however, a small percentage die of heart failure caused by infection of the heart valves.

# Prevention

Gonorrhea can be prevented by abstinence or by having sex only with a faithful partner who is free of disease. Condoms offer excellent protection. It is best to avoid oral sex altogether.

People who are being treated for gonorrhea should not have sex with anyone until they are completely cleared of infection. They should tell their partners so that they can be tested for gonorrhea and treated if necessary.

Babies are protected against the disease by having silver nitrate drops placed in the eyes to prevent eye infection.

Having had gonorrhea offers no protection at all against reinfection. There is also no vaccine against the disease.

## The Future

Gonorrhea is likely to continue to be a major public health problem for the foreseeable future for several reasons. One is the high rate of infection with other STDs among people diagnosed with gonorrhea. A second reason is complacency about the ease of treatment with older antibiotics; many people do not yet realize that drug-resistant strains of gonorrhea are increasingly widespread. A third reason is that many men, as well as women, are willing to have unprotected sex in exchange for alcohol or recreational drugs. In addition, the use of drugs and alcohol affects

# WORDS TO KNOW

- **Abstinence:** Not having sexual intercourse with anyone.
- **Carrier:** A person who has an infectious disease and can transmit it to others but has no symptoms of the disease themselves.

**Clap:** A slang term for gonorrhea.

Disseminated gonococcal infection (DGI):

A complication of gonorrhea in which the disease organisms get into the bloodstream and cause arthritis, eye disease, skin rashes, or inflammation of the heart valves.

**Ectopic pregnancy:** A pregnancy in which the fertilized egg starts growing outside the uterus, usually in the abdomen or in the tubes leading to the uterus.

- **Gonococcus:** The bacterium that causes gonorrhea.
- **Pelvic inflammatory disease (PID):** A complication of gonorrhea that can leave a woman permanently unable to have children.
- **Pus:** A body fluid produced in response to bacterial infections. It consists of dead white blood cells and tissue fluid.
- **Urethra:** The tube that allows urine to pass from the bladder to the outside of the body.

people's judgment about the health risks involved in sex with a large number of partners. Last, the Internet has made it increasingly easy for people to meet others for casual or risky sex.

SEE ALSO AIDS; Chlamydia; Genital herpes; Syphilis

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# Graves Disease

# Definition

Graves disease is an autoimmune disorder of the thyroid gland, which means that an abnormal immune response causes the person's thyroid gland to produce too much of thyroid hormone known as thyroxine. It is named for Robert James Graves (1797–1853), an Irish physician who first described a patient with goiter and exophthalmos in 1835.

## Description

In Graves disease, the patient's thyroid gland, which is located at the base of the throat, is stimulated by an autoimmune response to increase its production of thyroxine. Doctors do not know what triggers the body's immune system to target the thyroid gland, although some think that genetic factors may be involved. Other researchers think that bacterial or viral infections may trigger the onset of the disorder, as it often appears suddenly, particularly in middle-aged adults. Overproduction of the hormone then causes such characteristic symptoms of the disease as anxiety, general restlessness, sleep problems, bulging eyes, heat sensitivity, and weight loss.

# Demographics

Graves disease is the most common cause of hyperthyroidism in the United States, accounting for 70–80 percent of cases and affecting about

#### Also Known As

Hyperthyroidism, Basedow disease, diffuse toxic goiter

#### Cause

Autoimmune disorder of the thyroid gland

#### Symptoms

Exophthalmos (protruding eyes), goiter, fatigue, weight loss with increased appetite

#### Duration

Months to years



Bulging eyes on an elderly woman, a symptom of her Graves disease. SPL / PHOTO RESEARCHERS, INC.

thirty people in every 100,000. It appears to run in some families, with patients often having relatives diagnosed with either Graves disease itself or Hashimoto disease, another thyroid disorder. As of 2008, however, no specific gene had yet been identified as the sole cause of Graves disease.

The disorder is most common in adults over the age of twenty but may affect children as well. It is seven times as common in women than in men. Most affected women are between the ages of thirty and sixty.

Graves disease is found in all races and ethnic groups, but the genetic markers that have been linked to the disorder appear to vary with race and ethnicity.

# **Causes and Symptoms**

In Graves disease, a person's immune system mistakenly produces antibodies that target the thyroid gland; however, instead of destroying the gland, the antibodies stimulate it to make too much thyroxine. Some researchers believe that age and stress may combine with genetic factors to trigger the onset of the disease. In a few cases, Graves disease appears to be triggered by traumatic injury to the thyroid gland.

The most common symptoms of Graves disease are:

- Anxiety, irritable mood, and difficulty sleeping.
- Losing weight in spite of normal or even increased food intake.

# Graves Disease and the Bush Presidency

Graves disease is not a communicable disease, and it is rare for it to be diagnosed in a husband and wife within the space of two years. Nonetheless, in the spring and summer of 1991, President George H. W. Bush was diagnosed with Graves disease, only two years after First Lady Barbara Bush had been diagnosed with the same illness.

The President was out jogging one day in May 1991 when he felt a sudden tiredness, shortness of breath, and a rapid irregular heartbeat. He was taken to Bethesda Naval Hospital, where his doctors quickly determined that the problem was in his thyroid gland. He had developed a goiter, had lost nine pounds in the previous two months while eating normally, and had felt unusually tired for the previous two weeks. Bush chose to be treated with radioactive iodine and has taken a replacement thyroid medication ever since.

Barbara Bush had also been treated with radioactive iodine when she was diagnosed with Graves disease in April 1989. The odds that a husband and wife would independently develop Graves disease are thought to be about one in three million. As a result, the doctors who examined the Bushes began to question whether a virus might have been responsible for triggering the disorder in both spouses.

A specialist in thyroid disorders from the Mayo Clinic in Minnesota who was consulted about the case simply described it as a "bizarre coincidence"; it was the only explanation he could think of.



George H.W. Bush and Barbara Bush. AP IMAGES.

- Exophthalmos. This is a symptom in which the eyeballs bulge outward from the eye socket. This symptom develops because the tissues and muscles behind the eye swell and push the eyeball forward. It is more likely to develop in patients who smoke.
- Goiter. This symptom is an enlargement of the thyroid gland that causes a bulge at the base of the neck.
- Muscle weakness and tremor.
- Brittle hair.
- Increased sensitivity to heat and heavy sweating.
- Speeded-up or irregular heartbeat.
- In women, lighter than normal menstrual periods.
- Frequent bowel movements.
- Double vision and eye irritation.

Undiagnosed and untreated Graves disease can lead to a condition known as thyroid storm, in which the patient's body temperature rises as high as 105°F (40.5°C). Thyroid storm is a medical emergency and can end in death if not treated. The symptoms of thyroid storm include chest pain, heavy sweating, shortness of breath, disorientation, and very rapid heartbeat as well as a rise in body temperature. The patient should be taken to a hospital for emergency treatment.

Other complications that can result from untreated Graves disease include osteoporosis (brittle bones) and weakened heart muscle leading to congestive heart failure.

## Diagnosis

People should visit their doctor for a checkup if they notice such outward signs of Graves disease as goiter, weight loss, or increased sensitivity to heat. The doctor will begin by taking a history, including a family history of thyroid disorders. He or she will then check the patient's eyes for exophthalmos and feel the thyroid gland for swelling. The patient's blood pressure and pulse will also be checked.

The doctor will then order blood tests to check the levels of thyroxine in the patient's blood and another hormone known as thyroid-stimulating hormone or TSH. In Graves disease, the thyroid gland is stimulated by an abnormal antibody rather than by TSH, so a blood test that indicates high levels of thyroxine and low levels of TSH points to Graves disease as the cause.
Another test that is used to diagnose Graves disease consists of injecting the patient with radioactive iodine and measuring the amount that is taken up by the thyroid gland. The thyroid gland needs iodine to produce thyroxine, so that if the test shows that the gland is taking up the radioactive iodine in higher amounts, it indicates that the patient has an overactive gland.

### Treatment

There are three major forms of treatment for Graves disease: antithyroid drugs that lower the amount of thyroid hormone produced; radioactive iodine, which destroys all or part of the thyroid gland; and surgical removal of all or part of the thyroid gland. Antithyroid drugs are about 30–50 percent effective in relieving the symptoms of Graves disease and are generally not recommended for younger patients. Radioactive iodine and surgery are 90–95 percent effective, but patients must take replacement thyroid hormone for the remainder of their lives. Radioactive iodine is not recommended, however, for patients with severe eye problems associated with Graves disease.

#### Prognosis

The prognosis for recovery from Graves disease depends partly on the patient's overall health and partly on how quickly the disorder is diagnosed. Most patients have good results provided they take replacement thyroid as prescribed and see their doctors for regular follow-up visits. Patients are usually advised to watch for signs of fever, sore throat, and throat ulcers, and see their doctor at once if they develop those symptoms. In most cases of Graves disease, no matter which therapy is used, eventually the patient will wind up with an underactive gland and require replacement thyroid hormone.

## Prevention

There is no known way to prevent Graves disease.

## The Future

Graves disease is not likely to become more common in the future. It is likely, however, that researchers will discover more information about the genetic factors involved in the disease and the specific conditions that trigger its onset.

**SEE ALSO** Hashimoto disease; Lupus; Osteoporosis; Rheumatoid arthritis; Sjögren syndrome

## WORDS TO KNOW

Autoimmune disorder: A disorder characterized or caused by autoantibodies that attack the cells or organs of the organism producing them.

- Exophthalmos: Abnormal protrusion of the eyeballs.
- **Goiter:** A swelling in the neck caused by an enlarged thyroid gland.
- **Hyperthyroidism:** A disease condition in which the thyroid gland produces too much thyroid hormone.
- **Metabolism:** The chemical changes in living cells in which new materials are taken in and energy is provided for vital processes.
- **Thyroid storm:** A medical emergency marked by a rise in body temperature as well as other symptoms caused by untreated hyperthyroidism.

Thyroiditis: Inflammation of the thyroid gland.

**Tremor:** Trembling or shaking caused by a physical disease.

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## Gulf War Syndrome

## Definition

Gulf War syndrome (GWS) is an illness associated with combat veterans of the 1991 Gulf War. It is controversial because of the wide variety of symptoms claimed as its characteristic features and because of the lack of a standardized definition of it. A report issued by the Institute of Medicine (IOM) in 2006 concluded, "although veterans of the first Gulf War report significantly more symptoms of illness than soldiers of the same period who were not deployed, studies have found no cluster of symptoms that constitute a syndrome unique to Gulf War veterans."

## Description

GWS has been described in various ways: as an immune system disorder, a form of amyotrophic lateral sclerosis (Lou Gehrig's disease), a type of chronic fatigue syndrome, a skin disorder, a cause of birth defects in veterans' children, a disease of the digestive tract, a neurological disorder, or a lung disease. Specific symptoms include loss of energy and muscle control, headaches, dizziness and loss of balance, memory problems, difficulty concentrating, muscle and arthritis-like pains in the joints, indigestion, skin rashes, shortness of breath, and even insulin resistance (a condition that precedes diabetes). Other symptoms reported by veterans have included diarrhea, chest pain, irritability, sleep disorders, night sweats, bleeding gums, hair loss, and depression.

GWS was first reported in late 1991, shortly after veterans of the brief conflict began returning home. Many veterans began to complain of health problems that they thought were the result of service in the Persian Gulf. In 1998 Congress passed two laws directing the National Academy of Sciences to evaluate the medical and scientific evidence related to what the media had named Gulf War syndrome. The task was given to the Institute of Medicine, which has issued a series of reports on these health problems. It was the IOM's 2006 report that stated that there is no unique group or cluster of symptoms that can be identified as Gulf War syndrome.

## **Demographics**

About 700,000 American men and women served in the Gulf War in 1991; about 30 percent of them have registered with the database set up by the American Legion to track illnesses related to the war. Studies conducted by the Institute of Medicine and the National Academy of Sciences (NAS) surveyed 19,000 veterans of the Gulf War. Other medical reports by doctors in the field covered an additional 80,000 soldiers.

#### Also Known As

Gulf War illness, Persian Gulf syndrome, Saudi flu, desert fever

#### Cause

Disputed

#### Symptoms

Fatigue; headaches; memory and sleep problems; digestive, skin, muscle, and joint disorders

#### Duration

Unknown

## **Causes and Symptoms**

The causes of the symptoms associated with GWS are as controversial as the assortment of symptoms. Some that are still considered as possible causes as of 2006 include:

- Infectious diseases, particularly those caused by parasites carried by sandflies
- Combustion products from weapons that used radioactivity-depleted uranium
- Side effects from an anthrax vaccine that was used in the early 1990s
- Exposure to chemical weapons, particularly nerve gas and mustard gas
- Combinations of these causes

Other possible causes that have been eliminated since 1991 include:

- Toxins in the smoke from oil well fires
- Biological weapons
- Nitric acid compounds that were used in Iraqi rockets
- Multiple chemical sensitivity (MCS, another controversial condition that lacks an exact diagnosis)
- Overconsumption of aspartame, an artificial sweetener used in soft drinks

Psychological stress is still considered a major factor in causing the physical symptoms associated with GWS, however. The findings of the various IOM studies that identify stress as a cause of many of the symptoms are not accepted by everyone, however. As one epidemiologist explains, "This has been stigmatized—the idea that there may be stress-related illnesses. I think that there's still a certain perception among some people that for some reasons, stress and psychological illnesses aren't real, which is certainly not the case."

One reason why some people have difficulty accepting that psychological stress could cause the physical symptoms of GWS is that the war itself was very short in duration. Thus many assumed it was a low-stress war. This assumption was wrong, however, because the troop buildup took several months before actual combat began. In addition, the soldiers



Soldier being treated for Gulf War syndrome at Walter Reed Army Medical Center. © SCPHOTOS / ALAMY.

## Identifying a New Disease

Gulf War syndrome (GWS) is controversial. It is one of a series of war-related syndromes in American history that have never been clearly defined. Since the Civil War (1861–1865), American war veterans have reported a higher rate of stress-related physical and mental symptoms than their civilian counterparts. In 1871, physician J. M. Da Costa wrote about Civil War veterans describing what he called "irritable heart syndrome." This condition resembled what was later called shell shock in World War I (1914–1918), battle fatigue in World War II (1939–1945), and posttraumatic stress disorder (PTSD) in the Vietnam conflict (1954–1975).

A doctor at the Naval Medical Research Institute compared studies of the veterans of these wars and concluded there were two general categories of war-related illness, one physical and one psychological, but neither has been clearly defined based on laboratory tests or other precise measurements. The diagnosis of physical or psychiatric disorders depended on the soldier's own description of symptoms and the doctor's impression.

Studies aiming to identify GWS as a distinct illness are part of epidemiology. Epidemiologists track the frequency and spread of diseases—mental and physical—in large populations. They decide whether a "new" disease or disorder is a previously unrecognized condition or whether it is a different form of a known disease. They could not determine a standard set of diagnostic criteria that defines GWS. The lack of a definition is a major social and medical problem. Naming a disorder has a powerful effect on people's perception of the condition, the way doctors treat patients, and how patients understand their illness. The controversy surrounding GWS was intensified by media reports of a wide range of unusual health problems among veterans.

had heard rumors of chemical and biological weapons as well as the existence of Iraqi missiles; they could not know in advance that the war would be brief; and the desert climate was itself a stressful environment.

## Diagnosis

There is no standard diagnostic description of Gulf War syndrome. The findings from the studies carried out by the IOM and NAS showed that the majority of soldiers in the patient sample had illnesses that could be identified by current diagnostic criteria. In many cases, the soldiers would have had these health problems whether or not they had been sent to the Persian Gulf. The smaller group included veterans with the mix of physical and psychological symptoms associated with GWS. The important finding in regard to this smaller group is that their symptom cluster can also be found in about 10 percent of people in ordinary doctors' offices or health clinics. Thus, it is difficult to maintain that GWS is a distinctive syndrome when a considerable number of civilians report the same vague cluster of symptoms that do not point to any specific disease.

## Treatment

Treatment is based on the individual veteran's symptoms.

## Prognosis

The prognosis varies according to the individual veteran's symptoms.

## Prevention

There is no known way to prevent future illnesses that may be related to service in this particular war. The Institute of Medicine has noted that early reports of increased rates of cancer or birth defects among Gulf War veterans and their families are inconsistent. In addition, the IOM points out that cancer is a disease that can take years to develop, so that it will be some time in the future before epidemiologists can tell whether veterans of this particular war have increased rates of cancer compared to people of the same age who were not in combat.

## The Future

Epidemiologists have noted that unexplained symptoms among veterans are likely to recur after each war because humans still do not understand the complicated relationship between chronic but nonspecific symptoms and physical or emotional illness. In addition, researchers also do not understand all the social as well as medical factors that cause people civilians as well as combat veterans—to feel sick. Recommendations for the future include better physical and psychological screening of soldiers before as well as after combat as well as more extensive studies of chronic health problems in civilians as well as military personnel.

Another recommendation for the future concerns better communication between doctors studying war-related syndromes and the general public. One difficulty is the complexity of the medical research involved in tracking down any new disorder and defining its diagnosis. Explaining these complications requires a high degree of skill in preventing misunderstanding by listeners used to ten-second sound bites. Another common problem in communicating with the public is the uncertainty built into the practice of medicine. Many people think of medicine as a neat and tidy branch of science with no loose edges. Perhaps most important, however, is educating the public about a truth that many find hard to accept: that conditions like Gulf War syndrome are evidence of the human mind's ability to convert fear and other strong emotions into physical symptoms rather than diseases in the usual sense.

## WORDS TO KNOW

**Epidemiology:** The branch of medicine that deals with the frequency, distribution, and control of disease in a population.

Multiple chemical sensitivity (MCS): A controversial health condition related to a patient's belief that his or her symptoms are caused by exposure to environmental chemicals.

**Syndrome:** A group of signs or symptoms that occur together and characterize or define a particular disease or disorder.

SEE ALSO Chronic fatigue syndrome; Posttraumatic stress disorder

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## Gum Disease

See Periodontal disease.

# Η



## Genetic

Infection

Injury

Multiple

Other

Unknown

#### Also Known As

HFMD, enterovirus infection, coxsackievirus infection

Cause Virus

#### Symptoms

Fever, mouth ulcers, blisters on the hands, feet, and buttocks

#### Duration

A week to ten days



## Hand-Foot-and-Mouth Disease

## Definition

Hand-foot-and-mouth disease (HFMD) is a contagious virus infection most likely to affect children below the age of ten. Its most noticeable symptoms are sores in the mouth and a rash on the hands, soles of the feet, and sometimes the buttocks. HFMD should not be confused with foot-and-mouth (sometimes called hoof-and-mouth) disease, which is a virus infection that affects cattle, pigs, and sheep. Humans cannot get the animal disease, and they cannot transmit HFMD to household pets or other animals.

## Description

Hand-foot-and-mouth disease is largely a disease of young children in the United States. It often spreads rapidly in schools, day care centers, and other places where large numbers of children may be in close contact. The viruses that cause HFMD belong to a group of viruses called entero-viruses, which get their name from the fact that they are commonly found in the human digestive tract. The two most common enteroviruses that cause HFMD are called enterovirus 71 (EV71) and coxsackievirus A16, named for the town in New York where it was first identified in 1948.

The enteroviruses that cause HFMD are spread by contact with the mucus, tears, saliva, blister fluid, or feces of an infected person. The most

#### Hand-Foot-and-Mouth Disease

Child with a sore on the tongue from hand-foot-and-mouth disease. © HERCULES ROBINSON/ ALAMY.



common methods of transmission are contact with the unwashed hands of a person with HFMD or with a toy, drinking glass, or other object the infected person has touched. Infected children and adults are most contagious during the first week of illness; the virus can, however, remain in the intestines for about a month after the illness.

HFMD has an incubation period of three to seven days. The first symptoms of illness are usually fever and a sore throat, followed by loss of appetite. About two days after the fever begins, the patient develops painful sores in the mouth. A rash with blisters also appears on the palms of the hands, the soles of the feet, and sometimes on the buttocks or genitals. Some people have only the rash and some have only the mouth sores. The rash and mouth sores last for about a week or ten days and then clear completely.

Children and adults who have been infected with HFMD are immune to the specific virus that caused their symptoms after they recover. They can, however, develop a second case of HFMD if they are infected with a different enterovirus known to cause the disease.

## Demographics

Hand-foot-and-mouth disease is a common disease around the world, with millions of cases each year. It is primarily a disease of young

children, although young adults sometimes get infected. Older adults rarely get HFMD unless they have weakened immune systems.

There are epidemics of HFMD in the United States about every three years, most often in late summer or early fall. The disease affects both sexes and all racial and ethnic groups equally.

## **Causes and Symptoms**

Hand-foot-and-mouth disease is caused by at least fourteen different enteroviruses, the two most common in the United States being enterovirus 71 and coxsackievirus A16.

The most common symptoms of HFMD include:

- Low-grade fever (101°F [38.3°C])
- Sore throat
- Loss of appetite
- Painful reddish ulcers or blisters around or in the mouth or on the soles of the feet and palms of the hands
- Headache
- Tiring easily or sleeping more than usual
- Crankiness in infants and toddlers

Some children may also:

- Drool
- Develop muscle aches
- Have pains or cramping in the abdomen

## Diagnosis

The diagnosis of hand-foot-and-mouth disease is usually based on the doctor's observation of the patient's age, history, and visible symptoms. Blood tests or other laboratory studies are rarely done because the illness is usually mild. It is possible to identify the viruses that cause HFMD by taking stool samples or by swabbing the patient's mouth and throat and culturing the virus in the laboratory, but these tests can take between two and four weeks to yield results.

## Treatment

There is no specific medication that can cure hand-foot-and-mouth disease, as antibiotics are not effective against virus infections. Treatment is intended to ease the pain and discomfort of the fever, mouth sores, and other symptoms of HFMD. To bring down the fever and relieve muscle pain, children can be given acetaminophen, ibuprofen, or another nonaspirin fever reducer or pain reliever. The doctor may recommend a mouthwash, throat lozenge, or throat spray that contains a mild anesthetic to relieve pain caused by blisters inside the mouth or throat. Another treatment that can help to ease throat pain is to gargle with a salt water rinse made by adding half teaspoon of salt to an 8-ounce glass of warm water.

It is important to make sure that the patient drinks plenty of fluids. Children with HFMD sometimes become dehydrated because the sores inside the mouth and throat hurt when the child tries to drink fruit juice, soft drinks, tea, or other drinks that contain acid. Milk-based drinks or cold foods like ice cream or popsicles are often less painful for the child to swallow. Children who do become dehydrated may need to be taken to the hospital for treatment with intravenous fluids.

Children with blisters on the hands and feet should keep the areas clean by washing gently with soap and water, then patting the skin dry to avoid breaking the blister and spreading the infection. If the blister does open, it should be covered with a small bandage.

## Prognosis

Most people in any age group with HFMD recover completely in about a week. In a few cases, children have developed encephalitis, meningitis, or pneumonia; in rare cases, HFMD can cause death. An epidemic in Taiwan in 1998 that affected 1.5 million children resulted in 405 severe complications and seventy-eight deaths. An outbreak of HFMD in China that began in the spring of 2008 led to 25,000 reported cases and twenty-two deaths by early May.

Pregnant women who become infected with HFMD are at increased risk of losing the baby.

## Prevention

There is no vaccine against HFMD. The most effective preventive measure is frequent and thorough hand washing, particularly after using the toilet, changing a diaper, before meals, and before preparing food. Another preventive measure is routinely cleaning shared toys in day care centers as well as in the home with a disinfectant, because the viruses that cause HFMD can live on objects for several days. The Centers for Disease Control and Prevention (CDC) recommends washing toys (or soiled countertops and other surfaces) with soap and water, followed by using a solution of one tablespoon of chlorine bleach added to four cups of water.

Children with HFMD should stay home from child care or school until the fever has gone and the mouth sores and other blisters have healed. Adults with the disease should stay home from work.

## The Future

Hand-foot-and-mouth disease is likely to continue to be a common infection because there are so many different enteroviruses that can cause it, and because these viruses are the second most common family of viruses—only the viruses that cause the common cold are more widespread.

Researchers are presently working on a rapid diagnostic test that would allow doctors to distinguish quickly between coxsackievirus A16 and enterovirus 71 in patients with symptoms of HFMD. Such a test would be helpful during epidemics because EV71 infections are more likely to lead to complications than coxsackievirus infections.

SEE ALSO Encephalitis; Meningitis; Pneumonia

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## Hantavirus Infection

## Definition

Hantavirus infection is more commonly called hantavirus pulmonary syndrome or HPS in the United States. It is a rare but potentially fatal disease that people get by breathing in the virus from the dried urine, feces, or saliva of its vectors —infected mice or rats. Hantavirus infection is classified as both an emerging infectious disease and a zoonosis (disease transmitted from animals to humans).

## Description

The most distinctive symptom of hantavirus infection is its effect on the patient's lungs and breathing. After an incubation period of several days to several weeks, the infected person develops symptoms that can easily be mistaken for the flu or food poisoning. In the second phase, however, the patient has trouble breathing and may die as the lungs fill up with fluid and other body organs begin failing.

## Demographics

Anyone can get hantavirus infection from infected mice. However, most reported cases are in middle-aged adults. People of both sexes and all races can get the infection. Men are at somewhat greater risk than women, however, because they are more likely to be employed in occupations that expose them to infected mice. Hantavirus infection had been reported in thirty states in the United States as of 2008. It is most likely to occur in the spring and summer as the weather warms and people spend more time outdoors. It can also occur in the fall, when the rodents that carry the virus seek shelter from the cold weather in houses or barns.

Some people are at greater risk of hantavirus infection:

- People who live in New Mexico, Colorado, Arizona, California, Utah, Texas, Washington, Idaho, and Montana.
- Farmers, ranchers, and field hands
- Field biologists and veterinarians
- People who work in grain elevators, feed lots, or feed mills

#### Also Known As

Hantavirus pulmonary syndrome, HPS, Sin Nombre virus, SNV, Four Corners disease

## 

RNA virus

#### Symptoms

Fever, chills, headache, difficulty breathing, cough, death

**Duration** Four to ten days

- Utility workers, plumbers, and electricians
- Construction workers and building contractors
- Hikers and campers

## **Causes and Symptoms**

Hantaviruses are RNA viruses that enter the body when people breathe dust contaminated by the urine, feces, or saliva of rodents. They sometimes breathe in this contaminated dust while working in a mouse-infested building, cleaning out a barn



or shed, or hiking or camping in an area where infected mice build their burrows. When a person breathes in the dust, the virus particles enter the tissues lining the nose, throat, and lungs.

People can also take in the virus if they touch an object that has been contaminated by rodent droppings and then touch their mouth or nose. It may take anywhere from one week to five weeks for the disease to incubate. In a very few cases, people have been infected with hantavirus after being bitten by an infected mouse, but this method of transmission is unusual. As far as is known, people cannot get hantavirus infection through direct contact with an infected person.

The early stage of the disease is often misdiagnosed as flu or food poisoning because the person may have muscle ache and pains like those of flu or have nausea, vomiting, and diarrhea resembling the signs of food poisoning. This stage of the illness lasts from a few hours to several days. Other early symptoms of hantavirus infection are:

- Fever between 101 and 104°F (38 and 40°C)
- Chills
- A dry cough
- Rattling noises in the lungs
- Fatigue

In the second stage of the disease, the body responds to the virus that is infecting the tissue lining the lungs by secreting large amounts of fluid in order to get rid of the virus. The fluid, however, makes it hard for the person to breathe. Eventually the patient's blood pressure drops, an abnormal heart rhythm may develop, and one's breathing and blood circulation fails.

*Image of a hantavirus splitting.* © PHOTOTAKE INC. / ALAMY.

## **Discovery of an Emerging Disease**

The Indian Health Service and the CDC were puzzled by an outbreak of a new disease in the Four Corners region of the American Southwest in May 1993. A young, healthy Navajo man died shortly after having difficulty breathing. His fiancé had died several days earlier after having similar symptoms. Five other cases followed. The CDC sent special investigators to find the cause. Chief investigator Terry Yates (1950–2007) traced the disease organism to deer mice. An unusually large number of deer mice thrived in spring 1993 due to heavy rains that provided a bumper crop of food for the mice.

The cause of the deaths was a new virus in the hantavirus family called Sin Nombre virus or SNV. Since 1993, several other hantaviruses carried by different species of rats and mice in Florida, Louisiana, Rhode Island, and New York have been identified. The CDC also found that several mysterious deaths in the United States before 1993 had been caused by a hantavirus; the earliest known case concerned a Utah man in 1959.

The name "hantavirus" itself comes from the Hantaan River in South Korea, where the first virus in this family was discovered by researcher Dr. Lee Ho-Wang in 1976. Dr. Ho-Wang had been looking for the cause of a hemorrhagic fever that



Hantavirus researcher in New Mexico, during a 1996 outbreak. AP IMAGES.

had been killing thousands of people in Siberia, China, and Korea since the 1930s. About 2,300 American soldiers were infected by the hantavirus during the Korean War (1950–1953), and about 800 died as a result.

## Diagnosis

The diagnosis of hantavirus infection is based on the patients' history, including their occupational history or other evidence that they were exposed to mice and rats or their droppings. Hantavirus infection can be distinguished from influenza, bubonic plague, and other conditions that may cause painful breathing through a blood test. In most cases the doctor will also order a chest x ray to look for fluid buildup in the lungs.

## Treatment

There is no drug that can cure hantavirus infection. The patient's best chance of recovery is being placed on a respirator to help him or her breathe. Another form of treatment that helps some patients is extracorporeal membrane oxygenation, or ECMO. It is a technique in which the patient's blood is pumped through a machine that removes carbon dioxide and adds oxygen to the blood. The oxygenated blood is then returned to the patient's body.

## Prognosis

In the early 1990s hantavirus infection was almost always fatal. Better understanding of the disease, however, has reduced the death rate to about 50 percent. Patients who survive, however, recover completely without long-term damage to their lungs.

## Prevention

There is no vaccine to prevent hantavirus infection. The risk of hantavirus infection can be lowered considerably, however, by taking proper precautions to keep buildings free of rodents and to minimize exposure to them outdoors. The CDC recommends the following steps:

- Seal up holes, cracks, and other gaps that might allow mice to get into a house or barn.
- Get rid of any food sources (pet food, garbage cans, animal feed) within 100 feet (30 meters) of the house.
- Cut grass short near the house and keep shrubbery trimmed.
- Use mousetraps inside the house; disinfect dead rodents before disposing of them.
- Treat rodent droppings with chlorine bleach or another disinfectant before sweeping or vacuuming them. Wear rubber gloves during cleaning.
- Wear a respirator when cleaning, repairing, or working in a rodent-infested building.

## The Future

It is difficult to predict whether hantavirus infection will become more common in the United States and Canada over the next few decades. On the one hand, hantaviruses may spread to parts of North America where the infection has not yet been reported. On the other hand,

## WORDS TO KNOW

**Emerging infectious disease (EID):** A disease that has become more widespread around the world in the last twenty years and is expected to become more common in the future.

**RNA virus:** A virus whose genetic material is composed of ribonucleic acid (RNA) and

does not need DNA to copy itself and multiply.

**Vector:** An animal that carries a disease from one host to another.

**Zoonosis:** A disease that can be transmitted from animals to humans.

doctors are now better trained to recognize the disease early and hospitalize patients quickly. It is also possible that an effective vaccine will be developed at some future point.

SEE ALSO Influenza; Plague

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## Hashimoto Disease

## Definition

Hashimoto disease is an autoimmune disorder in which the body's immune system attacks the cells and tissues of the thyroid gland, a hormone-secreting (also called endocrine) gland resembling a butterfly located at the base of the neck just below the Adam's apple.

## Description

Hashimoto disease is caused by autoimmune damage to the thyroid gland at the base of the neck. The gland may become swollen—a condition known as goiter—or it may remain normal in size. In either case it becomes inflamed, and its cells begin to die. This loss of tissue means that the gland no longer produces enough thyroid hormone, a chemical that the body needs to regulate its metabolism. This condition is known as hypothyroidism.

## **Demographics**

Hashimoto disease is the most common cause of hypothyroidism in the United States and Canada in people over six years of age. It is largely a disease of adulthood, with the rate increasing with age. The most commonly affected age group is middle-aged adults between thirty and fifty.

Hashimoto disease is diagnosed in about fourteen women out of every 1,000 and one man in every 2,000. The disorder is estimated to be between ten and twenty times more common in women than in men, for reasons that are not yet known. It appears to be equally frequent in all races and ethnic groups, however.

## **Causes and Symptoms**

Although doctors know that Hashimoto disease is an autoimmune disorder, they do not know what triggers its onset. Some think that the Also Known As

Hashimoto thyroiditis, chronic lymphocytic thyroiditis

#### Cause

Autoimmune disorder in which the body's own cells attack the thyroid gland

#### Symptoms

Fatigue, constipation, dry skin, weight gain, cold intolerance, hair loss

#### Duration

Lifelong following diagnosis

#### Hashimoto Disease

Image of thyroid gland tissue affected by Hashimoto disease. The pink areas have been damaged by the disease. CNRI / PHOTO RESEARCHERS, INC.



disease is related to a gene known as HLA-DR5; however, different genes seem to be related to the appearance of the disease in different ethnic groups. It is known that the disease tends to run in families.

Other researchers think that a bacterium or virus may be the cause of the autoimmune response in Hashimoto's disease. In any case, the autoimmune processes cause the destruction of the cells in the thyroid gland, leading to a drop in the production of thyroid hormone and the characteristic symptoms of hypothyroidism.

The symptoms of Hashimoto disease are not specific to it; that is, they can be caused by other diseases or disorders. In addition, the symptoms typically come on gradually; the person may simply feel tired or less energetic than usual or develop dry, itchy skin and brittle hair that falls out easily. It may take months to years before the person or their doctor begins to suspect a problem with the thyroid gland.

Some patients with Hashimoto disease, however, have an early phase of the disease in which they have too much thyroid hormone in their bloodstream; this temporary hyperthyroidism is caused by the leaking of thyroid hormone from damaged cells in the gland. This condition is called hashitoxicosis and is characterized by anxiety, heavy sweating, restlessness, diarrhea, high blood pressure, and a general feeling of being keyed up. Eventually the damaged thyroid cells die, however, and the level of thyroid hormone in the blood drops below normal. Typical symptoms of Hashimoto disease include the following:

- Cold, dry skin and increased sensitivity to cold weather
- Dry brittle hair that falls out easily
- Constipation
- Hoarse voice and puffy face
- Unexplained weight gain of 10–20 pounds (4.5–9 kilograms), most of which is fluid
- Sore and aching muscles, most commonly in the shoulders and hips
- In women, extra-long menstrual periods or unusually heavy bleeding
- Weak leg muscles
- Memory loss
- Depression

## Diagnosis

The diagnosis of Hashimoto disease is usually made by tests of the patient's thyroid function. The first test is a hormone test for thyroid-stimulating hormone, or TSH. TSH is a hormone produced by the pituitary gland in the brain that stimulates the thyroid gland to produce thyroid hormone. When the thyroid gland is not producing enough hormone, the pituitary gland secretes more TSH; thus a high level of TSH in the blood indicates that the thyroid gland is not as active as it should be. Another type of

## Haraku Hashimoto (1881–1934)

Haraku Hashimoto was born into a family of medical doctors in the small village of Midau on the island of Honshu, Japan. He entered the new medical school at Kyushu University at the age of 22, graduating with one of its first classes in 1907. He intended to specialize in surgery, studying under Hayari Miyake (1867– 1945), the first Japanese neurosurgeon.

Hashimoto then went to Germany for postgraduate study. He published a paper in 1912 in a German medical journal on four cases of a disorder of the thyroid gland, noting the characteristic abnormalities of the gland's tissue that are still used in diagnosing the disease later named after him. Although Japanese doctors were unaware of Hashimoto's discovery, because it had been published in a German journal, English and American doctors who read the journal recognized that Hashimoto was describing a distinctive disorder, which they named Hashimoto's thyroiditis. Hashimoto himself continued to study in German and English hospitals until 1914, when his father died and he returned to Japan to take up his father's medical practice.

He specialized in major abdominal surgery after his return to Japan rather than continuing to work on disorders of the thyroid. He was only 53 when he died of typhoid fever in 1934.

blood test involves testing for the presence of abnormal antibodies. Because Hashimoto disease is an autoimmune disorder, there will be two or three types of anti-thyroid antibodies in the patient's blood in about 90 percent of cases.

The doctor may also order an ultrasound study of the patient's neck in order to evaluate the size of the thyroid gland or take a small sample of thyroid tissue in order to make sure that the gland is not cancerous. Thyroid tissue that has been affected by Hashimoto's disease has a distinctive pattern of broken cells and other types of tissue damage that will confirm the diagnosis.

## Treatment

Treatment for Hashimoto disease consists of a daily dose of a synthetic form of thyroid hormone known as levothyroxine, sold under the trade names of Synthroid, Levothroid, or Levoxyl. The patient is told that the drug must be taken as directed for the rest of his or her life.

In the early weeks of treatment, the patient will need to see the doctor every six to eight weeks to have their TSH level checked and the dose of medication adjusted. After the doctor is satisfied with the dosage level and the patient's overall health, checkups are done every six to twelve months. The reason for this careful measurement of the medication is that too much levothyroxine increases the risk of osteoporosis in later life or abnormal heart rhythms in the present.

## Prognosis

The prognosis for patients with Hashimoto disease is excellent, provided they take their medication as directed. They can usually live a normal life with a normal life expectancy.

The chief risks to health with Hashimoto disease are related to lack of treatment for the disorder. If this type of thyroiditis is not diagnosed and treated, patients are at increased risk of goiter, an enlarged heart, and severe depression. In addition, women with untreated Hashimoto disease have a higher risk of giving birth to babies with cleft palate and other birth defects.

## Prevention

There is no known way to prevent Hashimoto disease because the cause is not yet completely understood.

## The Future

The incidence of Hashimoto disease is not likely to increase in the foreseeable future. Research of the disorder is likely to focus on two questions: tracking down all the specific genes that may be involved in triggering the disorder; and relating Hashimoto disease to other autoimmune disorders that have a high female/male sex ratio. Some researchers think that there may be a common factor linking Hashimoto disease to

## WORDS TO KNOW

- Autoimmune disease: A disease in which the body's immune system attacks its own cells and tissues.
- **Goiter:** A swelling in the neck caused by an enlarged thyroid gland.
- Hashitoxicosis: A temporary phase in some patients with Hashimoto disease in which there is too much thyroid hormone in the blood due to leakage from damaged and dying cells in the thyroid gland.
- **Hyperthyroidism:** A disease condition in which the thyroid gland produces too much thyroid hormone.

- **Hypothyroidism:** A disease condition in which the thyroid gland does not produce enough thyroid hormone.
- **Levothyroxine:** The chemical name for the synthetic thyroid hormone given to treat Hashimoto disease.
- **Metabolism:** The chemical changes in living cells in which new materials are taken in and energy is provided for vital processes,

Thyroiditis: Inflammation of the thyroid gland.

lupus, rheumatoid arthritis, and other autoimmune disorders that disproportionately affect women.

SEE ALSO Graves disease; Hypothyroidism

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## **m** Hay Fever

## Definition

Hay fever is a form of allergic rhinitis, or inflammation of the soft tissues lining the nose. It is triggered by plant pollen, most commonly ragweed, tree, or grass pollen in the United States. Hay fever can be either seasonal, meaning that the person has symptoms only during certain periods of the year, or perennial, which means that the person has symptoms all year long. Children are more likely than adults to have perennial hay fever.

Hay fever is often grouped together with eczema and asthma as an atopic disease. Atopy is the medical term for the tendency to develop an allergy. About 20 percent of people with hay fever eventually develop asthma, and some develop eczema as well.

### Description

Hay fever is an allergic reaction to tree, grass, or weed pollen that is characterized by a stuffy or runny nose, sneezing, and teary or watery eyes. Some patients also complain of coughing, headaches, fatigue, and drowsiness. Although some patients have relatively mild symptoms that last for only a few weeks and are controlled by medications, others may suffer year round from symptoms and may develop such complications as ear infections or chronic inflammation of the sinuses. The National Institutes of Health (NIH) estimates that hay fever costs the United States about \$5.3 billion each year in terms of missed school and work days as well as the direct expenses of diagnosis and treatment.

### Demographics

It is estimated that between 20 and 25 percent of Americans have some form of hay fever, although the severity and the specific triggers vary

#### Also Known As

Allergic rhinitis, rose fever, grass fever

#### Cause

Allergic reaction to plant or tree pollen

#### Symptoms

Sneezing, itchy or runny eyes, stuffy nose, coughing

#### Duration

May be seasonal (a few months per year) or yearround

#### Hay Fever



Magnified image of ragweed pollen, a major cause of hay fever. © PHOTOTAKE INC. / ALAMY.

from region to region across the United States. It is difficult to determine whether various races or ethnic groups have different rates of sensitivity to pollen because of the variations in climate across the country and the different types of trees and grasses that are present. In general, the hay fever season is shorter in the northern states and longer in the South, particularly along the East Coast.

About 80 percent of people with hay fever develop it before age twenty, with the average age at onset being eight to eleven years. Some doctors think that as many as 40 percent of children may suffer from hay fever at some point in childhood. The symptoms typically become less severe in adult life and may go away completely in some cases.

Boys are more likely than girls to suffer from hay fever. Among adults, however, men and women are equally likely to have symptoms.

## **Causes and Symptoms**

The basic cause of hay fever is exposure to plant pollen. The most common sources are plants or trees that are pollinated by the wind rather than by bees or other insects. The reason for this difference is that windborne pollen particles are lighter and more likely to remain in the air than the heavier pollen grains produced by flowering plants that depend on insects to carry the pollen from one plant to another. For example, ragweed pollen has been found as high as 2 miles (3.2 kilometers) in the

## Tips for Minimizing Pollen Exposure

Although it is impossible to avoid airborne pollen completely, people who suffer from hay fever may be helped by the following suggestions:

- Check local news sources for the daily pollen count, and stay indoors during the early morning and evening hours; pollen levels are higher at those times.
- Use a face mask designed to filter out pollen if it is necessary to go outdoors.
- Do not dry clothes outdoors.
- Keep windows closed in the house and car; use the air conditioner to control temperature.
- Take a shower after outdoor activity; wash hair each night to remove pollen before going to bed.
- Minimize exposure to other substances that irritate the nasal passages, including cigarette smoke, chlorine bleach and other household chemicals, insect sprays, wet paint, and strong perfumes.
- Have someone who does not get hay fever do yard work.
- Avoid large fields and grassy areas if possible.
- Consider vacationing at the beach or on a cruise rather than in an inland area.

One approach that usually does not work, however, is relocating to a different part of the country. People who move to get away from a specific type of pollen often develop new allergies within a few years in the new location. atmosphere and as far as 400 miles (644 kilometers) out to sea. Common sources of airborne pollen include:

- Trees: birch, alder, willow, oak, ash, elm, poplar, olive, hazel, mountain cedar, and horse chestnut.
- Grasses: ryegrass, Kentucky bluegrass, redtop grass, Johnson grass, Bermuda grass, and timothy.
- Weeds: ragweed, sorrel, mugwort, sagebrush, plantain, and nettle.

Hot, dry, and windy days are more likely to trigger the symptoms of hay fever than cool or rainy days. The pollen count is also likelier to be higher early in the morning than later in the day.

When the pollen from these plants enters the patient's airway and the tissues that line the eyelids, immune system reacts to the pollen as an allergen, a substance that triggers an allergy. The process releases a compound called histamine. Histamine is responsible for the runny nose, itchy and watery eyes, and sneezing that characterize hay fever.

## Diagnosis

The diagnosis of hay fever is based on a combination of the patient's history and skin tests for specific allergens. It is sometimes difficult to tell at first whether a person has hay fever or the common cold, but in general, the symptoms of a cold get better in a few days or a week. When taking the patient's history, the doctor will ask when the symptoms began; whether the symptoms appear during the same time of the year; whether the symptoms are continuous or off and on; and whether they are worse at specific times of day. In some cases the patient has already noticed that certain activities or places seem to trigger the symptoms. To perform a skin test, the doctor takes a small amount of material extracted from a specific type of pollen and injects it under the patient's skin or applies it to a tiny scratch on the arm or upper back. If the patient is allergic to the material, his or her skin will develop a wheal, or flattopped reddish swelling. The skin test cannot be used on patients with eczema, however. These patients can be tested for plant allergies by a type of blood test called a radioallergosorbent test (RAST).

## Treatment

Treatment of hay fever has three parts: avoiding the specific trigger(s); using medications to relieve such symptoms as itching eyes and a runny nose; and taking allergy shots to reduce one's sensitivity to triggers.

Medications that are often recommended for hay fever include:

- Antihistamines. These are drugs that counteract the effects of histamine in causing the watery eyes and runny nose associated with hay fever. Some are available over the counter, while others require a prescription. The older antihistamines often make people drowsy. Thus, patients who use these should not drive or operate machinery while taking them. The newer antihistamines are more expensive but do not make people drowsy as a side effect.
- Decongestants. These are sprays or tablets that clear up congestion in the nose. Most can be purchased over the counter while others require a prescription. People with high blood pressure should avoid oral decongestants because they can raise blood pressure.
- Nasal corticosteroids. These are sprays that reduce the inflammation of tissues associated with hay fever. They may take about a week to start to work but are safe for long-term use.
- Gargling and rinsing out the nasal passages with salt water. There are over-the-counter nasal sprays available for his purpose, or patients can make their own rinse by adding one-quarter teaspoon of salt to two cups of warm water. Salt water works well to relieve nasal congestion in many people.

## Prognosis

The prognosis for hay fever varies. People with mild symptoms usually do well with a combination of antihistamines and decongestants used as needed during pollen season. Between 85 and 90 percent of patients who are treated by desensitization (allergy shots) benefit from this type of therapy. Some patients continue to do well after the shots are stopped but others find that their symptoms return with the next pollen season. In addition, desensitization carries some risk of a severe allergic reaction.

People with hay fever are at increased risk of chronic sinus disorders, nosebleeds, asthma, or ear infections.

## Prevention

The best way to prevent hay fever is to avoid things that trigger it. Some techniques for avoiding or minimizing exposure to ragweed and other pollens are listed in the sidebar. Another preventive approach is desensitization, which is also called immunotherapy. In desensitization, patients are given a series of injections of their specific allergen under the skin, with the concentration of allergen in the shots being gradually increased. It takes an average of eight to twelve months for the patient to see results, however, and the injections must be taken for at least three years and sometimes closer to five years.

### The Future

Hay fever appears to be increasing in frequency among adults in developed countries, although the reasons for the rise are not yet known. One theory is that adults who get hay fever for the first time were either not exposed to common allergens as children or that they become sensitized to allergens when their immune systems are weakened by a viral infection or pregnancy.

A newer form of treatment for hay fever that shows promise is a vaccine called Pollinex Quattro that was developed in the United Kingdom. Instead of receiving desensitization shots for three to five years, patients using the new vaccine get only four shots—one per week for a month. The vaccine was tested on patients in the United States and Canada as well as Europe in the summer of 2007. Researchers hope that the vaccine could be available as early as 2010 following further clinical trials.

SEE ALSO Asthma; Common cold; Eczema

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- **Allergen:** Any substance that causes an allergic reaction. Pollen is the allergen that triggers hay fever attacks.
- **Atopy:** The medical term for an allergic hypersensitivity that affects parts of the body that are not in direct contact with an allergen. Hay fever, eczema, and asthma are all atopic diseases.

Chronic: Long-term or recurrent.

**Desensitization:** A form of treatment for hay fever that involves a series of shots containing the

allergen to reduce the patient's sensitivity to that particular trigger. Desensitization is also called immunotherapy.

- **Histamine:** A compound that is released during an allergic reaction.
- **Rhinitis:** The medical term for inflammation of the mucous tissues lining the nose. It can be caused by infections or chemical irritants as well as by allergies.
- **Wheal:** A suddenly formed flat-topped swelling of the skin; a welt.

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## <mark>m</mark> Headache

## Definition

A headache is a disorder in which a person feels pain or discomfort somewhere in the face, neck, or scalp. The brain and the skull are not the sources of headache pain because they do not contain pain-sensitive nerve endings. The sources of headache pain are nerve endings in the scalp, face, throat, the muscles of the head, and blood vessels at the base of the brain. When any of these nerve endings are triggered by stress, tension in the muscles, inflammation, or dilation of the blood vessels in the head, the person may experience pain.

Doctors have described over 130 different types of headaches. They can be classified into primary and secondary headaches. Primary headaches are those in which the headache is not caused by an injury, infection, or other disorder, but rather by some type of disturbance in the brain's relationship to the body. They include migraine headaches, tension headaches, cluster headaches, and so-called ordinary headaches.

Secondary headaches are caused by an injury or some other illness. There are at least 300 known causes of secondary headaches. The major types of secondary headaches are post-traumatic headaches, sinus headaches, reactive headaches, and rebound headaches.

It is possible for a person to suffer from more than one type of headache.

## Description

Headache pain can vary in location, severity, duration, and quality (dull, piercing, throbbing, etc.) depending on the cause or type of the headache. The main characteristics of the major types of headaches are described below.

Primary headaches:

• Migraine headaches: Migraine headaches are caused by disturbances in the central nervous system leading to swelling of the blood vessels

#### Also Known As

Cephalagia, head pain

#### Cause

Muscle tension, infections, spasms in arteries around the brain, tumors, stroke, injury

#### Symptoms

Pain in various parts of the scalp, face, or neck; nausea and vomiting; visual disturbances

#### Duration

Minutes to days

Headache

in the brain and severe pain. The pain affects only one side of the head in 60 percent of cases, and is often accompanied by nausea, vomiting, and extreme sensitivity to light. There are two basic types of migraine: migraine with aura (visual disturbances preceding the pain of the headache) and migraine without aura. The person may be sick for one to two full days.

- Tension headaches: These are characterized by a sensation of tightness or pressure in the head and are often accompanied by muscle tension in the neck. Tension headaches may occur on a daily basis or only at random. They usually last for several hours.
- Cluster headaches: Cluster headaches are sharp and extremely painful headaches that tend to occur several times per day for months and then go away for long periods of time. They are the rarest type of primary headache.
- Ordinary headaches: Some doctors think that ordinary headaches are actually a mild form of migraine. These headaches usually occur at random, are not associated with a head injury or other illness, and usually go away with rest and mild pain relievers. Secondary headaches:

Secondary headaches:

• Post-traumatic headaches: Post-traumatic headaches occur in as many as 88 percent of people with a closed head injury and 60

## When to See the Doctor

Most headaches go away by themselves or with over-the-counter pain relievers in a few hours. Some headaches, however, indicate a serious health problem. Anyone with any of the following symptoms should see their doctor or go to the emergency room at once:

- The headache comes on suddenly and has a violent or explosive quality.
- The headache feels like the worst one the person has ever had.
- The person is experiencing slurred speech, change in vision, problems moving arms or legs, loss of balance, confusion, or memory loss along with the headache.
- The headache is getting worse over a twenty-four-hour period.
- The person has fever, stiff neck, nausea, and vomiting along with the headache.
- The person is over age fifty and the headaches just began.
- The person is losing consciousness or is having convulsions.
- The person suffered a head injury before the headache.
- The headache is so severe that it wakes the person from sleep.
- The headache has lasted longer than a few days.

percent of people with a whiplash injury. This type of headache is accompanied by pain in the neck and shoulders, dizziness, mood or personality changes, and sleep disturbances.

• Sinus headaches: These are associated with post-nasal drip, sore throat, and a discharge from the nose. The pain of a sinus head-ache is usually experienced in the front of the face and head, and is usually worse in the morning than later in the day.

- Reactive headaches: Reactive headaches are triggered by an irritant in the environment or another illness. There are hundreds of possible triggers, ranging from the weather, pollen, dust, and other allergens, to colds, flu, eyestrain, and stomach upsets.
- Rebound headaches: Rebound headaches are a reaction to overuse of over-the-counter medications for pain relief, decongestants, or muscle relaxants. They can also be caused by withdrawal from caffeine or alcohol.

## Demographics

Headaches are a very common problem in the general population. Almost everyone gets an occasional headache, particularly when they are short on sleep, emotionally stressed, have skipped a meal, or are suffering from flu or a cold. Children can get headaches as well as adults; by age six, 31 percent of children have had at least one headache; by the time a child is fifteen, the number has risen to 70 percent. Between 60 and 80 million Americans suffer from frequent headaches but only 30 percent of these people consult a doctor for treatment.

According to the National Institutes of Health (NIH), children in the United States miss 1 million days of school each year because of headaches while adults miss 160 million days of work. Headaches cost the economy an estimated \$30 billion each year in medical expenses.

Headaches affect people of all races equally; however, the gender ratio varies depending on the type of headache. Women are three times as likely as men to suffer from migraine headaches, but men are ten times as likely as women to get cluster headaches.

### **Causes and Symptoms**

The basic causes of headaches include disturbances in the central nervous system leading to irritation of the blood vessels in the head; tension in the muscles of the head and neck; infections; allergens and other environmental triggers; overuse of or withdrawal from drugs; lack of sleep; clenching or grinding the teeth; menstruation; depression or anxiety; certain foods; and head injuries.

Less common but dangerous causes of headaches include:

- Brain tumors
- Stroke

- An infection of the brain (encephalitis or meningitis)
- Bursting of a blood vessel in the brain

In addition to the pain of a headache, people may experience nausea, vomiting, diarrhea and other digestive symptoms; dizziness, loss of balance, and visual disturbances; mood and personality changes; extreme tiredness; muscle cramps in the neck and shoulders; inability to concentrate; and extreme sensitivity to light or noise.

## Diagnosis

Diagnosing headaches can be complicated because there are so many potential causes and because some people have more than one type of headache. In addition to examining the patient's head, neck, mouth, and throat in the office, most doctors will ask the patient to keep a headache diary, noting the time when a headache occurs, how long it lasts, other symptoms that accompany the headache, the quality and location of the pain, possible triggers, and other illnesses that the patient had at the time.

In some cases the doctor will order a computed tomography (CT) scan or a magnetic resonance imaging (MRI) of the patient's head. If encephalitis or meningitis are suspected, the doctor may order a spinal tap.

## Treatment

Treatment depends on the type of headache. Secondary headaches are treated by removing or avoiding the underlying cause, whether a head or whiplash injury, environmental trigger, food allergy, overuse of alcohol or medications, sinus infection, eyestrain, or other problem.

Primary headaches are usually treated by appropriate medications:

- Migraine headaches can be treated either by medications taken before an attack to stop it or reduce its severity, or by medications taken to relieve the headache after it begins. Preventive medications include a group of drugs called triptans; certain antidepressants; and antiepileptic drugs. After the headache starts, the patient may be treated with over-the-counter pain relievers like acetaminophen, naproxen, or ibuprofen, or prescription medications like ergotamine. Most patients with migraine are helped by resting in a quiet darkened room.
- Tension headaches: Usually respond well to over-the-counter pain relievers or to prescription pain relievers containing codeine. Hot showers and rest are also recommended for self-care at home. Some

patients are also helped by biofeedback, relaxation training, yoga, or massage therapy. In some cases the doctor may recommend psychotherapy if the patient's headaches are related to emotional stress.

- Cluster headaches: The triptans are effective in treating cluster headaches in many patients, as is oxygen inhalation. Because cluster headaches often come on very quickly, the triptans are usually given by injection rather than by mouth.
- Ordinary headaches: Usually treated in the same way as tension headaches.

## Prognosis

The prognosis for a headache depends on whether it is primary or secondary and its underlying cause or causes. Most ordinary headaches can be treated at home with few long-term side effects or complications. Cluster headaches, recurrent tension headaches, and migraines require long-term follow-up with a doctor. Cluster headaches are more difficult to treat successfully than either migraines or recurrent tension headaches.

## Prevention

People can lower their risk of headaches in several ways:

- Getting enough rest, eating a healthful diet without skipping meals, and exercising regularly.
- Taking occasional work or study breaks, particularly if working at a computer or reading for long periods of time.
- Having the eyes checked regularly, particularly if the person wears prescription eyeglasses or contacts.
- Avoiding overuse of over-the-counter pain relievers, decongestants, caffeine, or alcohol.
- Quitting smoking.
- Practicing relaxation techniques, yoga, meditation, or other approaches to stress management.
- Avoiding allergens, foods, or other factors known to trigger headaches whenever possible.

## The Future

Headaches are likely to be an ongoing health problem in the general population, if only because they have so many possible causes and

## WORDS TO KNOW

- **Aura:** A symptom that precedes migraine headaches in some people. The person may see flashing or zigzag lights, or have other visual disturbances.
- **Biofeedback:** An alternative treatment for headaches (and other conditions) that consists of teaching patients to consciously control their blood pressure, muscle tension, temperature, and other body processes.
- **Migraine:** A type of primary headache characterized by severe pain, nausea and vomiting, and sensitivity to light. It may occur on only one side of the head.
- Withdrawal: A collection of signs and symptoms that appear when a drug (including caffeine and nicotine) that a person has used for a long time is suddenly discontinued.

environmental triggers. Research into the causes of migraine headaches has yielded new insights since the late 1990s. Clinical trials include research into the causes of cluster headaches, which are still not well understood; evaluations of newer triptan drugs in treating migraine and cluster headaches; studies of the factors that affect the prognosis for recovery from headaches; studies comparing different types of treatment for rebound headaches; and studies of yoga, acupuncture, massage therapy, and other alternative treatments.

**SEE ALSO** Alcoholism; Allergies; Brain tumors; Common cold; Concussion; Encephalitis; Influenza; Meningitis; Stroke; Whiplash

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## m Hearing Loss

## Definition

Hearing loss is a disorder in which a person begins to lose the ability to hear in one or both ears. It may come on suddenly or develop slowly over a period of years; it may be temporary or permanent, and vary in severity from mild hearing loss to total deafness. There are many possible causes of hearing loss ranging from birth defects and ear infections (common causes in children) to exposure to high levels of noise in the workplace and the aging process (common causes in adults).

There are two major categories of hearing loss, defined by whether the loss results from problems in the structures of the outer or middle ear or whether it results from damage to the hair cells of the inner ear. The first type is called conductive hearing loss (CHL) and the second type is called sensorineural hearing loss (SNHL). CHL is often reversible while SNHL is not. People who have both CHL and SNHL are said to have mixed hearing loss.

## Description

Conductive hearing loss occurs when sound waves cannot move through the structures of the outer and middle ear. Ordinarily, sound

#### Also Known As

Deafness, hearing impairment, being hard of hearing

#### Cause

Head injuries, birth defects, infections, longterm noise exposure, medications, aging

#### Symptoms

Difficulty hearing conversations, radio, or television; difficulty learning to talk (children)

#### Duration

Days to years

#### Hearing Loss

Brain image of a patient with benign tumor (green circle) that results in progressive deafness. MEHAU KULYK / PHOTO RESEARCHERS, INC.



waves are funneled into the ear by the pinna, the visible part of the outer ear. The sound waves then pass through the ear canal, where they cause the eardrum and three tiny bones called ossicles to vibrate. The vibrations of the ossicles cause the liquid inside a snail-shaped structure
# Did You Know?

Doctors have found that unprotected exposure to sounds above 85–90 decibels (dB) for long periods of time can harm a person's hearing. Here is a list of the decibel levels of various common sounds:

- Weakest sound that can be heard: 0 dB.
- Soft whisper: 30 dB.
- Normal conversation: 60–70 dB.
- Piano music: 60-70 dB.
- Telephone dial tone: 80 dB.
- Freeway traffic: 85-90 dB.
- Trombone or French horn music: 90– 110 dB.
- Subway train 200 feet away: 95–100 dB.
- Power saw: 110 dB.
- Rock music: 115-120 dB.
- Pneumatic drill: 125 dB. This is the level at which most people feel pain in the ear.
- Gun blast, jet engine: 140 dB. This is the loudest recommended noise level even with hearing protection. Even shortterm exposure can cause permanent hearing loss.
- Loud rock concert: 150 dB.
- Death of nerve endings and other hearing-related tissues: 180 dB.
- Loudest possible sound: 194 dB.

called the cochlea to move. The movement of the liquid in turn causes hair cells inside the cochlea to respond. The hair cells convert movement into electrical signals that are then relayed to the brain via the auditory nerve. Conductive hearing loss can occur when the ear canal is blocked by wax or a foreign object, the ear drum is punctured, the ossicles are dislocated, or the ear canal is swollen shut due to infection.

Sensorineural hearing loss is caused by damage to the hair cells in the cochlea or to the nerves that conduct hearing signals to the brain. This damage can be caused by infections (measles, mumps, rubella, influenza, or mononucleosis); by trauma; by diabetes and other disorders that affect the circulatory system; by cancer drugs and some other medications; or by a tumor affecting the auditory nerve. SNHL is sometimes associated with such problems as tinnitus (ringing in the ears) or dizziness.

#### Demographics

Hearing loss is a common problem in the general American population, particularly in older adults. According to the Centers for Disease Control and Prevention (CDC), most people over the age of twenty begin to develop a mild hearing loss. A third of adults over the age of seventy have trouble hearing. Hearing loss is more common in older men than in older women.

About 24,000 children (three in every 1,000) are born with hearing loss in the United States each year. Causes include genetic disorders, infections before birth (particularly rubella), absence of ossicles or other abnormalities in the shape or inner structures of the ear, or low birth weight.

Hearing loss is equally common in all racial and ethnic groups, as far as is known.

#### **Causes and Symptoms**

The most common causes of conductive hearing loss are infections, trauma to the outer or middle ear, a buildup of earwax in the ear canal, foreign bodies in the ear, or dislocation of the ossicles caused by a blow to the ear.

The causes of sensorineural hearing loss include noise-induced hearing loss (NIHL), which causes trauma to the acoustic nerve; changes in atmospheric pressure inside the ear during deep-sea diving; fracture of the bone at the side and base of the skull; drugs that damage the nerves involved in hearing (cancer drugs, some antibiotics, diuretics, and aspirin or ibuprofen); diabetes; tumors on the auditory nerve; infectious diseases (mumps, measles, syphilis, meningitis, mononucleosis, and herpes); and aging.

The symptoms of hearing loss depend partly on the person's age. A baby who has not yet learned to talk or a child with hearing problems may have the following symptoms:

- Not responding to cooing or conversation from the parents or other family members
- Does not react to sudden loud noises
- Has trouble with certain word sounds
- · Does not repeat words or phrases used by others
- · Uses gestures to communicate with others
- Seems to watch people's faces for clues to understanding what they are saying
- Has trouble paying attention in school
- Turns up the radio or television louder than other members of the family

In adults, the symptoms of hearing loss may include:

- Problems hearing over the telephone
- Having trouble following conversations, particularly if two or more people are talking
- · Having to ask others to repeat what they have just said
- Having difficulty hearing higher-pitched sounds, such as the voices of women and children
- Failing to hear the doorbell or telephone ring
- Having difficulty telling the direction of a sound

#### Diagnosis

Diagnosing hearing problems in babies or toddlers is critical because the period from birth to three years of age is when children learn to use language. Hearing difficulties during this period can affect a child's ability to speak normally. To test hearing in infants and small children, an audiologist (hearing professional) can perform a variety of tests.

In adults, the doctor will examine the ear canal for signs of infection, a foreign object, or damage to the ear drum. A primary care doctor can test each ear separately with a tuning fork to check for conductive hearing loss, but the patient may be referred to an audiologist for more detailed measurement of the type and extent of hearing loss.

#### Treatment

Treatment for hearing loss depends on the cause. Infections of the outer and middle ear can be treated with medicated ear drops or oral antibiotics. Earwax and foreign bodies in the ear are removed by suction, forceps, or flushing the ear canal with water. If the earwax has hardened, the doctor may use special drops to soften it and have the patient return a few days later to have it removed. Hearing loss caused by medications is treated by discontinuing the medication.

A tumor of the auditory nerve will usually be removed by a neurosurgeon or an otolaryngologist (a doctor who specializes in ear, nose, and throat disorders). Patients with sensorineural hearing loss are also usually referred to ear, nose, and throat specialists for evaluation and treatment.

Patients whose hearing loss is caused by exposure to high levels of noise in their workplace will be advised to wear earplugs or other protective equipment. Well-fitted ear plugs can reduce noise level by about 25 dB. In extreme cases, the patient may be advised to switch jobs.

Conductive hearing loss can be treated by hearing aids, which are electronic devices that fit in or behind the ear and amplify sounds. A recent variation on traditional hearing aids is the bone-implanted hearing aid or BAHA. A BAHA is implanted in the patient's skull by a neurosurgeon. It consists of a titanium post that allows a sound processor to be attached outside the skull. The processor transmits sound waves to the titanium implant, which transfers the sound vibrations to the skull and inner ear. BAHAs are recommended for patients who cannot wear hearing aids inside the ear or for those with one-sided hearing loss. Another newer treatment for severe sensorineural hearing loss is the cochlear implant. A cochlear implant is an electronic device that is inserted in the inner ear by a surgeon and connected to a device worn outside the ear. Unlike a traditional hearing aid, a cochlear implant does not make sounds louder or clearer. Instead it works by stimulating the auditory nerve directly and bypassing damaged hair cells in the cochlea. Cochlear implants can be used only in adults or children over the age of twelve.

#### Prognosis

The prognosis of hearing loss depends on the cause and type. CHL is often reversible; typically, patients who suffer conductive hearing loss as a result of a plug of earwax or a foreign body in the ear, an infection of the outer or middle ear, or a ruptured eardrum will find that their hearing returns to normal after treatment.

Hearing loss caused by a medication may or may not improve after the drug is stopped. There is no proven treatment that can restore hearing other than discontinuing the drug.

Hearing loss caused by meningitis, tumors of the auditory nerve, and aging is usually permanent.

#### Prevention

Hereditary hearing loss cannot be prevented, but there are ways that other people can lower their risk of hearing loss as they get older:

- Avoid using several noisy machines at the same time.
- Learn to enjoy music, television, or radio programs at a moderate sound level.
- Avoid going to loud rock concerts on a frequent basis. Listening to rock music is a common cause of sensorineural hearing loss in teenagers and young adults.
- Wear earplugs when operating noisy equipment or when exposed to loud background noise for long periods of time. Earplugs can mean the difference between a safe and a dangerous level of noise.
- If work or commuting involves exposure to high noise levels, choose quiet activities for recreation or leisure time.
- See a doctor if hearing is lost suddenly or if there is pain, dizziness, or ringing in the ears.

#### WORDS TO KNOW

Audiologist: A health care professional who is specially trained to evaluate hearing disorders.	electrical signals that travel to the brain via the auditory nerve.
<b>Cochlea:</b> A snail-shaped fluid-filled chamber in the inner ear.	<b>Ossicles:</b> A group of three small bones in the middle ear that transmit sound waves to the
<b>Decibel (dB):</b> A unit of measurement for expressing the relative intensity of sounds.	cochlea.  Pinna: The visible part of the outer ear.
Hair cells: Special cells in the cochlea that convert the movement of the fluid inside the cochlea into	<b>Tinnitus:</b> The medical term for ringing in the ears.

## The Future

Hearing loss is a growing concern to public health doctors because there is evidence that it is a growing problem in the United States, particularly among younger adults. One study completed in 2008 estimated that as many as 29 million Americans have at least partial hearing loss.

SEE ALSO Brain tumors; Ear infection; Measles; Rubella

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# m

# Heart Attack

# Definition

A heart attack, also called a myocardial infarction or MI, is a potentially fatal health crisis caused by a loss of blood supply to the heart muscle. If normal blood flow is not restored within a few minutes, the tissue begins to die from lack of oxygen. Treatment should be started as soon as possible to prevent permanent damage to the heart.

Heart attacks are not the same thing as heart failure. Heart failure is a condition in which the heart cannot pump enough blood to meet the needs of the rest of the body. It usually develops slowly over a period of years and produces early symptoms like loss of energy or fluid buildup in the feet and ankles rather than sudden chest pain. A heart attack can, however, lead to heart failure.

# Description

The classic symptoms of a heart attack are pain in the chest, shortness of breath, nausea, and breaking out in a cold sweat. The patient may feel the pain as pressure or squeezing, a sensation of fullness or tightness, a heavy weight on the chest, or a mild or strong ache in the center of the Also Known As Myocardial infarction, MI

#### Cause

Death of heart tissue due to loss of blood supply

#### Symptoms

Chest pain, shortness of breath, sweating, nausea

**Duration** Several hours

#### Heart Attack

Image showing a blockage in a coronary artery, which resulted in a heart attack. JAMES CAVALLINI / PHOTO RESEARCHERS, INC.



chest. The pain may move from one part of the body to another, or extend from the chest to the jaw, arms, neck, or back. The person may also feel dizzy or lightheaded. The pain lasts for twenty minutes or longer, or it goes away briefly and then returns.

Not everyone with an MI has these classic symptoms. Some people may have a tight feeling only in the arms or upper back, feel mild indigestion or clammy skin, or may have trouble breathing. Women, people with diabetes, and the elderly are more likely than men to have mild or vague symptoms that can be easily missed; these are sometimes called silent heart attacks. Silent heart attacks are particularly dangerous because they are easy to ignore.

## **Demographics**

Heart attacks primarily affect adults. About 1.5 million Americans have heart attacks each year, and about half of them die within a year. Coronary artery disease, the major cause of heart attacks, is the leading killer of both men and women in the United States. Heart attacks are increasing worldwide, including the developing countries.

In the United States, heart attacks affect all races and ethnic groups equally.

Risk factors for heart attacks include:

- Lifestyle issues. People who smoke, consume large amounts of alcohol, or are physically inactive are at increased risk of heart attacks.
- Cocaine use. Cocaine causes blood vessels to tighten, thus potentially cutting off blood supply to the heart. Heart attacks in young adults are often caused by cocaine abuse.
- Family history of heart disease.
- Age. The risk of a heart attack increases after age sixty.
- Sex. Men are more likely to have heart attacks than women up to age seventy, when both sexes have an equal risk.
- High blood pressure and high blood cholesterol levels.
- Obesity.
- Diabetes.
- High levels of emotional stress.

## **Causes and Symptoms**

Heart attacks are caused by the loss of blood supply to the heart muscle, a condition known as ischemia. In about 90 percent of cases, blockage of the arteries that carry blood to the What to Do about a Heart Attack

A person having a heart attack needs to get help *as soon as possible*. If someone is having the symptoms of a heart attack:

- Call 911 within five minutes if possible.
- Call the doctor even if the symptoms go away in five minutes.
- Call for an ambulance to go to the hospital; do not take a private car because that will delay treatment.
- Take a nitroglycerin pill (or give one to the patient) if the doctor has prescribed this type of medicine.

Many people put off seeking help when they notice the warning signs because they are embarrassed to ask for help, do not want to cause trouble for other family members, or do not think their symptoms are serious. Doctors agree that it is much better to go to the hospital and find out that the symptoms have another cause than to wait too long and risk dying of a heart attack. According to the National Heart, Lung, and Blood Institute (NHLBI), about half the people who die from heart attacks die within an hour of their first symptoms and before they reach the hospital.

heart results from atherosclerosis, hardening of the arteries due to the formation of plaques along the walls of the blood vessel. Plaques are composed of a fatty material made up of dead white cells and cholesterol. If a plaque in one of the arteries supplying the heart ruptures, it can cause a blood clot to form in the artery and block it, thus starving the heart of blood.

The remaining 10 percent of heart attacks are caused by sudden spasms in the coronary arteries that shut down the flow of blood to the heart muscle. These spasms may result from cocaine use, a sudden emotional shock, or an abnormality in the shape of the coronary artery. The major symptoms of a heart attack are:

- Pain in the chest, which may be experienced as an ache, tightness, weight, or a squeezing sensation. The pain may move to the back, neck, arms, or jaw
- Nausea and vomiting
- Shortness of breath
- · Breaking out in a heavy cold sweat

Other symptoms of a heart attack may include lightheadedness or dizziness, intense anxiety, coughing, or a feeling that the heart is racing.

# Diagnosis

Most people having a heart attack will be taken to a hospital emergency room, where they will be asked to describe their symptoms. These questions help the doctor to rule out panic disorder, which is a type of anxiety disorder that can cause people to think they are having a heart attack. In addition to taking the patient's personal and family history of risk factors for a heart attack, the doctor will also take the patient's temperature, blood pressure, and pulse. Listening to the patient's lungs and heartbeat through a stethoscope can help to rule out pneumonia or other diseases that might cause chest pain or difficulty breathing.

The next step is diagnostic tests, which include:

- Electrocardiogram (ECG or EKG). An ECG or EKG measures the heart's electrical activity. Injured heart muscle makes unusual patterns or tracings on the paper printout produced by the ECG machine. If only a small amount of the heart muscle has been affected, the ECG may not show any abnormal patterns.
- Blood tests. These are done to confirm the diagnosis of a heart attack or to make sure that the electrocardiogram did not miss a small heart attack. Injured heart muscle leaks small amounts of enzymes into the bloodstream, which can be detected in a blood test. The emergency room doctor may repeat this blood test after several hours because it takes time for these enzymes to show up in the patient's blood.
- Chest x ray. A chest x ray may be done to see whether the patient's lungs are normal.
- Coronary angiography. Coronary angiography is a type of x-ray study in which the doctor threads a long thin tube called a catheter

into the heart through an artery in the arm or upper thigh. A dye that will show up on x ray is injected into the bloodstream through the catheter. This test allows the doctor to find the location of the blockage in the coronary artery.

#### Treatment

Treatment of a heart attack begins before the diagnosis is confirmed. The emergency room doctor will give the patient oxygen to help with breathing, aspirin to prevent further blood clotting, nitroglycerin to speed up the blood flow through the coronary arteries, and morphine or another pain reliever to make the patient comfortable.

The next step is the administration of clot-busting and bloodthinning drugs. These drugs can improve the patient's chances of survival and reduce the long-term damage to the heart. The patient may also be given beta-blockers, a group of drugs that slow down the heart rate and lower blood pressure; statins, drugs that lower blood cholesterol levels; or medicines to treat abnormal heart rhythms, which often develop after a heart attack.

In some cases the patient may need surgery. The two operations that are most commonly performed are coronary artery bypass surgery and coronary angioplasty. In bypass surgery, the surgeon takes a piece of a healthy artery from another part of the patient's body and sews it in place to go around a blocked coronary artery. This procedure will restore normal blood flow to the heart. In a coronary angioplasty, the surgeon inserts a catheter with a special balloon tip into the coronary artery. When the catheter is in the proper position, the balloon is expanded, which reopens the blocked artery. The surgeon will then insert a stent, which is a tube made of metal mesh, to keep the artery open.

Patients who survive their heart attack usually undergo rehabilitation after they leave the hospital. Rehabilitation includes lifestyle changes and psychological counseling as well as medications to keep the heart healthy.

#### Prognosis

The prognosis for recovery from a heart attack depends on how quickly the patient is diagnosed and treated as well as his or her age and overall health. About 30 percent of people do not survive their first heart attack; another 5–10 percent die within a year after the event. About half of patients diagnosed with a heart attack will need to be rehospitalized within a year.

A person who lives through the first two hours after the attack is likely to survive but may have complications like heart failure or blood clots in the lungs. Patients who do not have complications may recover completely.

# Prevention

People who have already had a heart attack can lower their risk of a second by taking a daily aspirin, other blood-thinning medications, cholesterol-lowering medications, beta-blockers, or other drugs that the doctor may prescribe to lower the strain on their heart muscle.

People who have not yet had a heart attack can lower their risk by:

- Quitting smoking or not starting in the first place.
- Getting regular medical checkups. This precaution is important because risk factors for heart attacks like high blood pressure, high cholesterol levels, and diabetes have no symptoms in their early stages.
- Avoiding using cocaine and drinking large quantities of alcohol.
- Keeping one's weight at a healthy level and getting regular exercise.
- Controlling blood pressure.
- Learning how to manage emotional stress.
- Eating a low-fat diet rich in fruits and vegetables.

## The Future

People are much more likely to survive heart attacks than they were in the 1960s because of the introduction of clot-busting drugs and improvements in heart surgery. New drugs to treat heart attacks are currently being studied as well as the effectiveness of using bone marrow or stem cells to help repair injured heart tissue.

**SEE ALSO** Coronary artery disease; Hypercholesterolemia; Hypertension; Panic disorder; Stroke

# For more information

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# WORDS TO KNOW

- **Atherosclerosis:** Stiffening or hardening of the arteries caused by the formation of plaques within the arteries.
- **Cholesterol:** A fatty substance produced naturally by the body that is found in the membranes of all body cells and is carried by the blood.
- **Ischemia:** Loss of blood supply to a tissue or organ resulting from the blockage of a blood vessel.

- **Plaque:** A deposit of cholesterol along the inside wall of an artery.
- **Stem cell:** A type of unspecialized cell that can reproduce itself and differentiate into different types of specialized cells. Stem cells act as a repair system for the body.

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# <mark>m</mark> Heart Diseases

Heart diseases are a group of disorders that affect the heart's ability to pump enough oxygenated blood to the rest of the body to meet its needs. Some heart diseases are caused by birth defects that slow down the flow of blood or change the path of the blood's flow between the heart and the lungs. Other heart diseases are caused by infectious diseases like rheumatic fever, which can cause an inflammation of the valves in the heart. Still others are caused by lifestyle choices like alcoholism, smoking, or failure to get treated for such conditions as diabetes or high blood pressure.

Heart diseases may involve damage to the heart muscle itself, damage to the valves inside the heart, or damage to the arteries that supply the heart. Partial or complete blockage of one of these arteries is the most common heart disease in the United States and can lead to a heart attack.

Heart diseases are the leading cause of death in North America and a major cause of disability. Almost 700,000 people die of heart disease in the United States each year, according to the Centers for Disease Control and Prevention (CDC). That figure represents 29 percent of all American deaths.

**SEE ALSO** Congenital heart disease; Coronary artery disease; Heart attack; Heart failure; Hypercholesterolemia; Hypertension; Rheumatic fever

# **m** Heart Failure

# Definition

Heart failure is a condition in which the heart cannot pump enough blood to meet the body's needs. A healthy heart can pump out 60 percent of the blood it receives in one beat; a failing heart pumps only 40 percent or less. Heart failure is not the same thing as a heart attack or cardiac arrest. Heart failure may develop either suddenly, in which case it is called acute heart failure, or slowly over a period of time, in which case it is called chronic heart failure.

Though most cases of heart failure involve both sides of the heart, it can be classified as left-sided or right-sided heart failure, depending on which side of the heart is affected. Left-sided heart failure is more common. In left-sided heart failure, the heart cannot pump enough oxygenated blood from the lungs to the rest of the body, leading to fluid buildup in the lungs. This buildup is called congestion, which is why heart failure is sometimes called congestive heart failure. The patient typically feels short of breath with left-sided heart failure. He or she may tire easily with even small amounts of exercise and have trouble breathing at night when lying flat.

In right-sided heart failure, the heart does not pump enough blood to the lungs to be oxygenated. As a result, fluid may collect in the patient's feet, ankles, and abdomen, causing swelling in the feet and ankles. In some cases the liver also becomes enlarged, and the veins in the patient's neck swell up.

#### Description

Heart failure can occur in children or adolescents but is usually a disorder of adults. In most cases the symptoms develop slowly over a period of months and years and are often attributed to aging. As the heart muscle gradually weakens—often as the result of a disease like diabetes or longterm high blood pressure, damage caused by a heart attack, or a congenital abnormality of one of the heart valves—the heart works harder to meet the body's needs for the oxygen and nutrients carried by the blood. As the heart becomes less efficient, the person often feels tired or lacking in energy. Heart failure is often not diagnosed until the person begins to develop fluid buildup in their feet or legs, lungs, abdomen, or liver.

As the heart muscle is weakened, the heart tries to make up for its loss of strength in one or more of three ways. It may enlarge, which allows it to fill with more blood and so have more blood to pump to other parts of the body. Second, it may acquire more muscle mass, which allows it to pump blood more forcefully, at least for a time. Third, the heart may simply speed up and pump faster.

In addition to the heart's attempts to make up for its growing weakness, the body may also respond, either by narrowing its veins and arteries in order to maintain blood pressure, or by redirecting blood away Also Known As Congestive heart failure, cardiac failure, CHF

#### Cause

Inability of the heart to pump enough blood to supply the rest of the body

#### Symptoms

Swelling of feet, ankles, or abdomen; shortness of breath; weakness; rapid or irregular heartbeat

**Duration** Months to years

#### Heart Failure

X ray showing congestive heart failure. The heart is enlarged, and there is excess fluid in the lungs. © PHOTOTAKE INC. / ALAMY.



from less vital parts of the body to the brain and heart, which are the most vital organs. These responses help to explain why some people can go on for years without being aware that their heart has lost some of its ability to function.

#### Demographics

Heart failure is a common disorder in the general American population, particularly among older adults. According to the Centers for Disease Control and Prevention (CDC), about 5 million people in the United States were living with heart failure in 2008, with about 550,000 new cases diagnosed annually. More than 287,000 people die each year from heart failure in the United States. The disorder costs the country \$30 billion each year in direct health care costs.

Heart failure is more common among people over 65 than among younger adults. It is the most common reason for hospitalization for patients on Medicare. Among children, congenital (inborn) heart defects are the most common reason for heart failure.

Other risk factors for heart failure include:

• Sex. Men are more likely than women to develop heart failure; however, among adults over the age of 75, more women than men have the condition.

- Race. African Americans are more likely than members of other races to develop heart failure, to develop it at younger ages, to get worse faster, and to die from heart failure.
- Obesity. Excess weight puts a strain on the heart muscle.
- Diabetes. Diabetes increases a person's risk of coronary artery disease and high blood pressure.
- History of coronary artery disease (narrowing of the arteries) or high blood pressure. Coronary artery disease lowers the supply of oxygen to the heart muscle.
- Virus infections that may have weakened the heart.
- Heart attack. A heart attack weakens the heart's ability to pump blood. According to the CDC, 22 percent of men with heart attacks and 46 percent of women will develop heart failure within six years of the heart attack.
- Alcohol abuse. Too much alcohol can weaken the heart muscle.
- Sleep apnea. Sleep apnea lowers the supply of oxygen to the blood during the person's sleep time and increases the risk of developing irregular heart rhythms as well as weakening the heart muscle.
- Kidney disease. Disorders of the kidneys increase the risk of heart failure because they lead to fluid retention and high blood pressure.

# **Causes and Symptoms**

The causes of heart failure include a number of factors that can weaken the heart's ability to pump blood, ranging from congenital defects in the structure of the heart to infections, lifestyle choices, or other diseases and disorders in later life.

The most common symptoms of heart failure are:

- Shortness of breath. The person may have trouble sleeping unless propped up on pillows, or may wake up suddenly feeling short of breath.
- Persistent coughing or wheezing, or coughing up bloody mucus. This symptom is caused by fluid building up in the lungs.
- Swelling of the feet, ankles, or abdomen. The patient may gain several pounds of weight very suddenly or notice that their shoes feel tight.

- Tiredness and fatigue. The person may find that even minor tasks or chores, such as shopping or carrying a small bag of groceries, leave them unusually tired.
- Nausea and loss of appetite. Fluid building up in the abdomen affects the digestive tract, causing the person to feel full or sick.
- Memory loss and confusion.
- Rapid heartbeat. The patient may notice that the heart is beating faster and experience it as a racing or throbbing sensation.
- Need to urinate at night. In some people with swollen feet or ankles, the body is able to dispose of some of the fluid at night through the urine.

# Diagnosis

The diagnosis of heart failure is complicated because many of the symptoms of the disorder are not unique. The doctor will usually begin with the patient's history and note such risk factors as a previous heart attack, diabetes, or high blood pressure. The doctor will then listen to the patient's heart and lungs with a stethoscope to detect evidence of congestion in the lungs or abnormal heart sounds.

If the doctor suspects that the patient has heart failure, he or she will order one or more laboratory or imaging tests:

- Blood test. This may be done to rule out kidney disease as the cause of fluid retention or to test for the presence of a hormone that is found in the blood when the heart is overworked.
- Electrocardiogram (ECG). This test measures the electrical activity of the heart.
- Chest x ray. This imaging test can identify fluid in the lungs and enlargement of the heart.
- Echocardiogram. This is an important test that uses sound waves to produce an image of the heart on a video monitor. It can be used to measure the percentage of blood pumped out by the left ventricle—the heart's main pumping chamber—with each beat.
- Stress tests. In these tests, the patient is either asked to exercise on a treadmill or is given a medication that stresses the heart to determine whether there are blockages in the heart's arteries.
- Computed tomography (CT) or magnetic resonance imaging (MRI) scans of the heart.

• Cardiac catheterization. In this type of test, the doctor inserts a thin tube called a catheter into a blood vessel in the groin or arm and threads it through the aorta into the coronary arteries. Radioactive dye injected through the catheter makes the arteries and the left ventricle of the heart visible on an x ray.

# Treatment

Except for cases of heart failure caused by damaged heart valves (which can be corrected by surgery), heart failure cannot be cured but only controlled. Patients may be given one or more medications or surgical treatments to control their symptoms and prevent further damage to the heart.

Medications that may be prescribed to treat heart failure include:

- Diuretics. Sometimes called water pills, these are drugs that help the body get rid of excess fluid through the urine.
- ACE inhibitors. These are medications that lower blood pressure, improve blood flow, and decrease the workload on the heart.
- Digoxin. Also known as digitalis, this drug increases the strength of the heart's contractions and slows down the heartbeat.
- Beta blockers. These medications slow heart rate, lower blood pressure, and reduce the risk of abnormal heart rhythms.
- Aldosterone antagonists. These drugs enable the body to get rid of salt and water through the urine, which lowers the volume of blood that the heart must pump.

Patients with acute heart failure may require treatment in a hospital. Hospital care usually includes oxygen therapy and medications (most commonly diuretics and drugs to relax the blood vessels) given intravenously.

Severe heart failure that cannot be controlled by medications requires surgical treatment:

- Implantable cardioverter defibrillator (ICD). ICDs are devices that surgeons implant beneath the skin and attach to the heart with small wires. They monitor the heart rate and correct heart rhythms that are too fast.
- Cardiac resynchronization therapy (CRT). In this type of treatment, a pacemaker sends timed electrical impulses to both ventricles of the heart to coordinate their rhythm.
- Heart pump. A heart pump, sometimes called a left ventricular assist device or LVAD, is a device implanted in the abdomen and

attached to a weakened heart to help it pump blood more efficiently. Originally used to keep candidates for heart transplants alive while they waited for a donor heart, LVADs are now thought of as alternatives to transplantation for some patients.

• Heart transplant.

#### Prognosis

The prognosis of heart failure depends on the person's age, sex, race, lifestyle, and other diseases they may have that affect the heart. Heart failure usually shortens a person's life expectancy by several years. Between 5 and 20 percent of people hospitalized for acute heart failure die in the hospital.

#### Prevention

Some causes of heart failure, such as congenital malformations, cannot be prevented. People can, however, lower their risk of heart failure in adult life by watching their weight, avoiding heavy drinking or the use of illegal drugs, getting regular exercise, and eating a diet focused on fruits, vegetables, whole grains, low-fat diary products, and lean meat.

People being treated for diabetes, high blood pressure, or coronary artery disease can lower their risk of heart failure by taking all medications prescribed by their doctor, following their doctor's recommendations about diet and exercise, and having regular checkups.

#### The Future

Heart failure is expected to continue to be a common disease of older adults in developed countries because of increasing life expectancy. Until the 1990s, doctors focused on controlling patients' symptoms. More recently, however, doctors are recommending preventive health care and lifestyle changes in the early adult years, before people develop the symptoms of heart failure or other disorders that increase the risk of heart failure.

**SEE ALSO** Coronary artery disease; Diabetes; Heart attack; Hypertension; Sleep apnea

# For more information

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# WORDS TO KNOW

**Cardiac arrest:** Heart attack; a condition in which the circulation of the blood stops abruptly because the heart stops beating.

Congenital: Present at birth.

Ventricles: The two lower chambers of the heart.

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# Heat Cramps

## Definition

Heat cramps are the mildest of the three forms of heat illness that can develop when the body is exposed to heat. They are defined as brief, involuntary painful muscle spasms in the legs or other parts of the body involved in work or exercise outdoors in hot weather.

# Description

Heat cramps are painful but brief muscle cramps that occur during exercise or work in a hot environment. The muscles may twitch or jerk involuntarily. The cramping sensations may also be delayed and occur a few hours after the work or exercise.

# Demographics

Heat cramps can affect people of all ages who are not used to hot weather, are not drinking enough fluid, sweat heavily, or have not been properly conditioned (improved their level of physical fitness). The cramps are most likely to affect the parts of the body involved in heavy work, such as the calves, thighs, shoulders, and upper arms.

# **Causes and Symptoms**

Heat cramps result when a person sweats heavily during work or exercise in hot weather. Sweating is the body's way of regulating its internal temperature to get rid of heat. As sweat evaporates, it cools the body. In addition to losing water through sweating, however, the body also loses electrolytes, which are minerals that are necessary for the body to function properly. When the levels of sodium and other electrolytes in the blood fall too low, the painful sensations of heat cramps occur.

Conditioning (improved physical fitness) reduces the risk of heat cramps by increasing blood volume; causing people to sweat more quickly, which helps the body get rid of heat; and making the sweat more dilute, so that fewer electrolytes are lost from the body in the sweat.

## Diagnosis

Diagnosis of heat cramps is usually based on their characteristics: the cramps are painful; they are involuntary; they come and go; they are brief; and they usually go away on their own. There are no blood tests or other diagnostic studies that can detect heat cramps.

# Treatment

Heat cramps are not usually considered a serious health problem even though the muscle cramps may be temporarily painful. They can be treated at home by stopping exercise or work; resting for a few minutes; and drinking fluids mixed with salt to replace the fluids and electrolytes lost through perspiration. People can have either a sports drink like

#### Also Known As

Heat-related muscle cramps

#### Cause

Heat exposure followed by loss of water and minerals through sweating

#### Symptoms

Painful muscle spasms in the legs, arms, or other muscles involved in exercise

#### Duration

A few minutes

Gatorade or clear fruit juice, or mix their own salt solution by adding one-fourth to one-half teaspoon of table salt to a quart of water. Salt tablets should not be taken because they upset the stomach.

To ease the cramping sensations, a person can practice gentle stretching or range-of-motion exercises to relax the muscles, or gently massage the affected parts of the body.

A doctor should be consulted when:

- The muscle cramps last longer than an hour.
- The affected person cannot drink the needed fluids because of nausea and vomiting.
- The person has more serious symptoms of heat-related illness, including dizziness, headache, shortness of breath, extreme tiredness, and a temperature higher than 104°F (40°C).

The doctor may administer intravenous fluids and check the affected person for signs of heat exhaustion or heat stroke.



Catherine Ndereba of Kenya suffering from heat cramps after winning the women's 2004 Boston Marathon. AP IMAGES.

# Prognosis

Heat cramps usually go away by themselves once the person has cooled off and replaced fluids lost through sweating.

## Prevention

Preventing heat cramps is largely a matter of taking time to adjust to hot weather or visiting a hot climate and dressing sensibly for local weather conditions:

 Most people in temperate climates need time to acclimate, or adjust to seasonal temperature changes. People should work up gradually to outdoor activities during the first few warm days of summer rather than overdoing it. The same is true of visiting a country with a tropical or hot climate. It is best to keep one's activity level moderate for a few days rather than crowding in too many activities. It can take people between seven and fourteen days to adjust to a hot climate. Marathon runners generally take two weeks to acclimate to training in the heat.

- Wear loose-fitting and light-colored clothing; choose fabrics that absorb sweat, such as cotton; wear a hat outdoors.
- Drink some fluids before exercising or working outside in hot weather. The American College of Sports Medicine recommends drinking about 20 ounces (0.6 liter) of water or a sports drink two to three hours prior to exercise, and 10 ounces (0.3 liter) of water or a sports drink ten to twenty minutes before exercise. It is important to *not* use thirst as a guide to fluid intake; a person can become dehydrated before feeling thirsty enough to want a drink.
- Use sunscreen generously, as sunburn lowers the body's ability to get rid of excess heat.
- Avoid caffeinated beverages and alcohol; they cause the body to lose additional fluid through the urine.
- Exercise during the early morning or late evening, when the temperature is cooler and the humidity lower.
- Consult a heat stress index like the one printed in the American Council on Exercise fact sheet listed below to help decide whether it is safe to exercise outdoors. There are times when the heat and humidity are so high that exercise should be avoided. Heat cramps are likely to occur when the heat stress index (the apparent temperature) is between 90–105°F (32–40.5°C).

# **The Future**

Heat cramps are a common consequence of exercising or working outdoors without proper conditioning or precautions. They are not dangerous by themselves, however. They can be prevented by dressing appropriately for hot weather, drinking enough fluids, and consulting the local heat index before outdoor activity.

SEE ALSO Heat exhaustion; Heat stroke

# For more information

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# WORDS TO KNOW

- Acclimation: The process of adjusting to seasonal climate changes or to a new climate.
- **Conditioning:** The process of becoming physically fit through a program of diet, exercise, and rest.
- **Electrolytes:** Minerals that are essential for proper body functioning. They include potassium, sodium, calcium, and magnesium.
- **Heat illness:** A general term for heat-related disorders, ranging from heat cramps (the mildest) to heat stroke (the most serious).

Involuntary: Not under the control of the will.

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## Definition

Heat exhaustion is a condition in which the body is overwhelmed by exercising or working in a hot environment; it produces more heat than it can get rid of through evaporation of sweat or moving into cooler surroundings. Heat exhaustion is the intermediate form of heat-related illness, heat cramps being the mildest and heat stroke the most serious.

# Description

Heat exhaustion is characterized by thirst, headaches, muscle cramps, shortness of breath, and nausea. Most patients with heat exhaustion have a normal level of alertness, although some people become slightly confused or feel anxious.

# Demographics

Heat exhaustion is the most common form of heat-related illness seen by physicians, although the exact number of people affected every year is not known because people can be treated for heat exhaustion outside a hospital or doctor's office. According to the Centers for Disease Control and Prevention (CDC), Arizona has the highest rate of cases of heat exhaustion in the United States.

Heat exhaustion affects people from all races and ethnic groups. It affects males and females equally.

Some groups of people have a greater risk of heat exhaustion:

- Newborn infants. A baby cannot adjust to changes in temperature as efficiently as an adult can. In addition, babies have only a limited ability to get out of a hot environment.
- Elderly people. As with infants, the bodies of elderly people do not regulate internal temperature as effectively as those of younger adults. In addition, elderly people may have underlying illnesses that make them more vulnerable to heat stress.
- Workers whose jobs require working outdoors in hot weather or near ovens, blast furnaces, or other sources of heat.
- People who are not physically fit or have not undergone conditioning to get their bodies used to work or exercise in the heat.
- People who take certain types of medications, including diuretics, drugs that regulate blood pressure, tranquilizers, antihistamines, and drugs given to treat people with schizophrenia.
- Homeless people.
- Obese people.

## **Causes and Symptoms**

Like heat cramps, heat exhaustion is caused by the loss of water and salt from the body due to sweating during exposure to heat or vigorous physical exercise in hot conditions. High humidity makes it harder for the

#### Also Known As

Heat illness, heat stress, hyperthermia

#### Cause

Overworking the body in hot and humid weather

#### Symptoms

Heavy sweating, fainting, nausea and vomiting, headache, dark urine

#### Duration

Two to three hours with treatment

body to regulate its internal temperature through sweating, which is its normal way to get rid of heat when the outside temperature is 95°F ( $35^{\circ}$ C) or higher. As sweat evaporates, it carries body heat with it. In addition to losing water through sweating, however, the body also loses electrolytes, which are minerals that are necessary to proper body functioning.

Other factors that can affect the body's ability to regulate its temperature in hot, humid weather include drinking alcohol, which leads to losing more water through the urine, and wearing tight clothes or clothes made of fabrics that do not allow sweat to evaporate easily.

The symptoms of heat exhaustion are more severe than those of heat cramps; they may come on either gradually or suddenly. People suffering from heat exhaustion may feel dizzy and faint as a result of the loss of body fluids and minerals.

- Skin is hot and moist; the person may develop goose bumps.
- Body temperature may be normal or a few degrees above normal.
- Nausea and vomiting.
- Rapid heartbeat and weak pulse.
- Blood pressure is low or drops lower if the person tries to stand up.
- Patient's legs may be swollen.
- Urine is darker than normal.

#### Diagnosis

In most cases the diagnosis is obvious from the weather conditions and the person's level of activity before feeling ill. People can take care of heat exhaustion themselves by moving into a cooler location; by drinking cool (not cold) water or sports drinks; and by lying down with the legs propped on a pillow or cushion to raise them above heart level.

If the person does not feel better in about half an hour; if they start to lose consciousness; or if their temperature goes above 104°F (40°C), they should be taken to an emergency room as soon as possible.

#### Treatment

The most important aspect of treating heat exhaustion is to keep it from getting worse. Untreated heat exhaustion can develop into heat stroke, which is a much more serious condition. In some cases a doctor may give the patient intravenous fluids if he or she appears to be severely dehydrated and is vomiting or otherwise unable to take fluids by mouth.

## Prognosis

Most people recover from heat exhaustion within two to three hours with no long-term effects.

## Prevention

Preventing heat exhaustion is largely a matter of taking time to adjust to hot weather or visiting a hot climate and dressing sensibly for local weather conditions:

- Most people in temperate climates need time to acclimate, or adjust, to seasonal temperature changes. People should work up gradually to outdoor activities during the first few warm days of summer rather than overdoing. The same is true of visiting a country with a tropical or hot climate; it is best to keep one's activity level moderate for a few days rather than crowding in too many activities. It can take people between seven and fourteen days to adjust to a hot climate; marathon runners generally take two weeks to acclimate to training in the heat.
- Wear loose-fitting and light-colored clothing; choose fabrics that absorb sweat, such as cotton; wear a hat outdoors.
- Drink some fluids before exercising or working outside in hot weather. The American College of Sports Medicine recommends drinking about 20 ounces (0.6 liter) of water or sports drink two to three hours prior to exercise, and 10 ounces (0.3 liter) of water or a sports drink ten to twenty minutes before exercise. It is important to *not* use thirst as a guide to fluid intake; a person can become dehydrated before feeling thirsty enough to want a drink.
- Use sunscreen generously, as sunburn reduces the body's ability to get rid of excess heat.
- Avoid caffeinated beverages and alcohol; they cause the body to lose additional fluid through the urine.
- People who must take prescription medications for allergies, high blood pressure, heart conditions, or certain types of mental disorders should ask their doctor whether any of their medications affect their response to heat.

# WORDS TO KNOW

- Acclimation: The process of adjusting to seasonal climate changes or to a new climate.
- **Conditioning:** The process of becoming physically fit through a program of diet, exercise, and rest.
- **Electrolytes:** Minerals that are essential for proper body functioning. They include potassium, sodium, calcium, and magnesium.
- **Heat illness:** A general term for heat-related disorders, ranging from heat cramps (the mildest) to heat stroke (the most serious).
- Exercise during the early morning or late evening, when the temperature is cooler and the humidity lower. Workers in occupations that require them to work in hot environments are often encouraged to take rest breaks during periods of hot weather. Some companies also provide rest areas where workers can cool off.
- Consult a heat stress index like the one printed in the American Council on Exercise fact sheet listed below or the National Weather Service's heat index to help decide whether it is safe to exercise outdoors. There are times when the heat and humidity are so high that exercise should be avoided. Heat exhaustion is likely to occur when the heat stress index (the apparent temperature) is over 105°F (40.5°C).
- People with elderly friends or relatives should check on them during summer heat waves to make sure that they are in good health. Heat waves that last longer than two days put the elderly at risk of heat exhaustion.

# **The Future**

Heat exhaustion is a common hot-weather health problem or a consequence of exercising or working outdoors without proper conditioning or precautions. It is dangerous only if it progresses to heat stroke, however. Heat exhaustion can be prevented by dressing appropriately for hot weather, drinking enough fluids, consulting the local heat index before outdoor activity, and knowing when to slow down and cool off.

SEE ALSO Heat cramps; Heat stroke

# For more information

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# Heat Stroke

## Definition

Heat stroke is the most severe of the three forms of heat-related illness. In heat stroke, a person's body temperature rises to 104°F (40°C) or higher. Unlike heat cramps and heat exhaustion, however, heat stroke

#### Heat Stroke



is a life-threatening condition. It has two forms: exertional heat stroke (EHS), related to work or exercise in the heat; and nonexertional heat stroke (NEHS), which is not caused by working or exercising outside and primarily affects the elderly, chronically ill persons, and infants during heat waves.

#### Description

Heat stroke is a medical emergency that develops when a person's body can no longer get rid of excess heat through sweating and evaporation of the sweat. As a result, the body's core temperature rises, damaging the proteins and cell membranes in the body tissues and leading to organ failure, destruction of muscle tissue, the collapse of the cardiovascular system, and eventually death.

#### Demographics

According to the CDC, over 8,000 people died in the United States from heat-related illness between 1979 and 2003. People over the age of sixty-five account for 44 percent of heat-related deaths.

Heat stroke affects people from all races and ethnic groups. Men and women are equally affected by heat stroke; however, men are twice as likely as women to die from heat stroke because more men than women Pakistani child being treated for heat stroke at a local hospital during a 2005 heat wave. AP IMAGES.

Also Known As Hyperthermia

#### Cause

Exposure to hot weather and high humidity

#### Symptoms

Body temperature of 104°F (40°C) or higher; hot, dry skin; nausea; loss of consciousness

**Duration** Minutes to hours

# First Aid for Heat Stroke

The Centers for Disease Control and Prevention (CDC) give the following instructions for treating someone with heat stroke:

- Have someone call 911 while the person is being cooled as rapidly as possible.
- Depending on what is available nearby, the affected person can be cooled by putting him or her in a tub with cool water; spraying the person with a garden hose; putting the person in the shower and running cool water over them; or sponging the person with a damp cloth dipped in cool water.
- Take the person's temperature from time to time if a thermometer is available, and keep cooling them until it drops to 101–102°F (38.3–38.8°C).
- Call a hospital for further instructions if rescue workers are delayed.
- Do not give the person water to drink.
- If possible, loosen or remove some of the person's clothing.

are employed in occupations that require working outdoors in hot weather.

Some groups of people have a greater risk of heat stroke:

- Newborn infants. The body of a baby cannot adjust to changes in temperature as efficiently as an adult's. In addition, babies have a limited ability to exit a hot environment.
- Elderly people. As with infants, the bodies of elderly people do not regulate internal temperature as effectively as those of younger adults. In addition, elderly people may have underlying illnesses or take medications that make them more vulnerable to heat stress.
- Workers whose jobs require working outdoors in hot weather or near ovens, blast furnaces, or other sources of heat.
- People who are not physically fit or have not undergone a conditioning program to get their bodies used to work or exercise in the heat.
- People who take certain types of medications, including diuretics, drugs that regu-

late blood pressure, tranquilizers, antihistamines, and drugs given to treat people with schizophrenia.

- Homeless people.
- Obese people.

# **Causes and Symptoms**

Like heat exhaustion, heat stroke is caused by the loss of water and salt from the body due to sweating during exposure to heat or vigorous physical exercise in hot conditions. High humidity makes it harder for the body to regulate its internal temperature through sweating, which is its normal way to get rid of heat when the outside temperature is 95°F (35°C) or higher. As sweat evaporates, it carries body heat with it. In addition to losing water through sweating, however, the body also loses electrolytes, which are minerals that are necessary to proper body functioning. In heat stroke, the body's cooling mechanisms are overwhelmed, and the body's internal temperature starts to rise uncontrollably.

Other factors that can impair the body's ability to regulate its temperature in hot, humid weather include drinking alcohol, which leads to losing more water through the urine, and wearing tight clothes or clothes made of fabrics that do not allow sweat to evaporate easily.

Heat stroke is often preceded by the symptoms of heat exhaustion, which include nausea and vomiting, headache, muscle cramps, dizziness, and difficulty breathing. The symptoms of heat stroke itself usually include:

- Hot, flushed, dry skin
- Changes in level of consciousness, including hallucinations, confusion, and irrational behavior
- Rapid heartbeat, sometimes as high as 130 beats per minute
- Rapid, shallow breathing
- Blood pressure may be either normal or low
- Body temperature above 104°F (40°C) or rectal temperature above 106°F (41.1°C).

## Diagnosis

The diagnosis of heat stroke is usually obvious from the patient's situation and previous activities. In addition to taking the patient's temperature, doctors in the emergency room may also take a urine sample to check kidney function or a blood sample to check the level of the patient's electrolytes and blood sugar. A blood test can also be used to evaluate whether the patient's liver has been damaged. In addition to these laboratory tests, the doctor may also order a muscle function test to see whether the patient's muscle tissue has begun to break down.

## Treatment

Immediate treatment for heat stroke is essential as death or permanent brain damage can occur within minutes. Emergency treatment is focused on cooling the patient as quickly as possible to a core body temperature of 102°F (38.9°C). Cooling may be done by spraying water on the body, covering the patient with sheets soaked in ice water, or placing ice packs in the patient's armpits and groin area. The patient's temperature is not lowered further because they may start to shiver, and shivering will raise their internal temperature again.

If the patient is conscious, they may be given additional oxygen to breathe and intravenous fluids to restore their blood volume. In most cases these fluids will contain sugar in order to lower the risk of liver failure. Patients who are having muscle cramps or convulsions are usually given benzodiazepine tranquilizers, which relax the muscles and reduce the risk of damage to muscle tissue.

The patient will be kept in the hospital for at least forty-eight hours after emergency treatment and monitored for brain damage, signs of liver failure, or other complications. This period of observation is necessary because heat stroke can damage almost all major body systems.

# Prognosis

Although people have survived body temperatures as high as 114.8°F (46°C), any temperature above 106°F (41.1°C) is potentially fatal. People who receive prompt treatment for heat stroke have a 90 percent chance of survival; without prompt treatment, 80 percent will die.

## Prevention

Heat stroke is largely preventable by taking time to adjust to hot weather and dressing sensibly for local weather conditions:

- Most people in temperate climates need time to acclimate to seasonal temperature changes. People should work up gradually to sports and other outdoor activities during the first few warm days of summer rather than overdoing. The same is true of visiting a country with a tropical or hot climate; it is best to keep one's activity level moderate for a few days rather than crowding in too many activities. It can take people between seven and fourteen days to adjust to a hot climate; marathon runners generally take two weeks to acclimate to training in the heat.
- Wear loose-fitting and light-colored clothing; choose fabrics that absorb sweat, such as cotton; wear a hat outdoors.
- Drink fluids before exercising or working outside in hot weather. The American College of Sports Medicine recommends drinking about 20 ounces (0.6 liter) of water or sports drink two to three hours prior to exercise, and 10 ounces (0.3 liter) of water or a

sports drink ten to twenty minutes before exercise. Do *not* use thirst as a guide to fluid intake; a person can become dehydrated before feeling thirsty enough to want a drink.

- Use sunscreen generously, as sunburn lowers the body's ability to get rid of excess heat.
- Avoid caffeinated beverages and alcohol; they cause the body to lose additional fluid through the urine.
- People who must take prescription medications for allergies, high blood pressure, heart conditions, or certain types of mental disorders should ask their doctor whether any of their medications affect their response to hot weather.
- Exercise during the early morning or late evening, when the temperature is cooler and the humidity lower. Workers in occupations that require them to work in hot environments should take rest breaks during periods of hot weather. Some companies also provide rest areas where workers can cool off.
- Consult a heat stress index like the one printed in the American Council on Exercise fact sheet listed below or the National Weather Service's heat index to help decide whether it is safe to exercise outdoors. There are times when the heat and humidity are so high that exercise should be avoided. Heat stroke is likely to occur when the heat stress index (the apparent temperature) is over 105°F (40.5°C) and the person is exposed to it for a long period of time; if the heat index is 130°F (54.4°C) or higher, heat stroke is highly likely even with short exposure.
- People with elderly friends or relatives should check on them during summer heat waves. Heat waves that last longer than two days put the elderly at risk of heat exhaustion.
- People who do not have air conditioning in their homes should go to a library, shopping mall, or other public building that is air-conditioned during a heat wave. Even a few hours in a cooler location can help to lower the risk of heat stroke.

# The Future

Heat stroke is a common hot-weather disorder; it is often a consequence of exercising or working outdoors without proper conditioning or precautions. Heat stroke can be prevented in normally healthy individuals by

# WORDS TO KNOW

Acclimation: The process of adjusting to seasonal climate changes or to a new climate.

**Conditioning:** The process of becoming physically fit through a program of diet, exercise, and rest.

- **Electrolytes:** Minerals that are essential for proper body functioning. They include potassium, sodium, calcium, and magnesium.
- **Heat illness:** A general term for heat-related disorders, ranging from heat cramps (the mildest) to heat stroke (the most serious).

dressing appropriately for hot weather, drinking enough fluids, consulting the local heat index before outdoor activity, and knowing when to slow down and cool off.

It is possible that heat stroke may become more common in some parts of the United States in the summer time because of the growing size of the elderly population and others who do not tolerate heat well because of chronic illness. One problem is geography: most parts of the United States have uncomfortably high temperatures for at least part of the summer, and some areas have temperatures at or above 90°F (32.2°C) for weeks on end. In addition, large cities tend to be hotter than the surrounding areas.

#### SEE ALSO Heat cramps; Heat exhaustion

# For more information

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# Hemophilia

# Definition

Hemophilia is the name of a group of hereditary blood disorders characterized by deficiencies in the blood's ability to form clots. Although hemophilia varies in severity from person to person, all patients with the disease bruise easily and bleed for abnormally long periods of time when cut.

There are two major forms of hemophilia: hemophilia A, sometimes called classic hemophilia, which accounts for about 80 percent of cases; and hemophilia B, called Christmas disease, which accounts for the remaining 20 percent. Both types are caused by gene mutations, hemophilia A by a mutation of the F8 gene and hemophilia B by a mutation of the F9 gene. Both genes are located on the X chromosome, which means that females (who have two X chromosomes) can transmit the mutations that cause hemophilia, but males (who have only one X chromosome) get the disease.

There is a very rare form of hemophilia called acquired hemophilia, which means that the disease is not genetic but develops later in life. It results from an autoimmune reaction in which the body attacks its own production of coagulation factor VIII, one of the blood factors required for normal clotting.

#### Also Known As

Bleeder's disease, royal disease, Christmas disease

#### Cause

Genetic mutations on the X chromosome; in rare cases, autoimmune disorders

#### Symptoms

Prolonged bleeding from minor cuts; excessive bruising; nosebleeds; blood in urine or stool

Duration Lifelong
#### Hemophilia

Bruised leg of a hemophiliac, a week after falling from a bike. DR P. MARAZZI / PHOTO RESEARCHERS, INC.



## Description

Hemophilia is a disease that has been known for centuries, although ancient doctors could do little to treat it. It was not until 1803 that John Otto, a doctor in Philadelphia, noted that the disease ran in families but that only males suffered from it. Hemophilia became known as the royal disease in the later nineteenth century, when several descendants of Queen Victoria (1819–1901)—including the queen's youngest son, Leopold—died young from brain hemorrhages. Two of Victoria's daughters were carriers of the defective F8 gene and passed on the disease to the royal houses of Spain, Russia, and Germany.

It was not until the twentieth century that doctors were able to understand the cause of hemophilia. At first they thought that it resulted from unusually fragile blood vessels. In the 1920s, doctors thought that defective platelets, cells in the blood involved in clot formation, were to blame. By 1937, however, it was found that substances dissolved in blood plasma, the liquid part of blood, were a necessary part of the normal clotting process. These proteins in the plasma were called coagulation factors. By 1944, a doctor in Argentina found that there are two distinct forms of hemophilia, each caused by a deficiency of a specific coagulation factor. It was not until 1965, however, that another doctor discovered a way to separate the protein factors from the liquid part of blood plasma by a freeze-drying process. This method made it possible for people with hemophilia to be treated without frequent high-volume blood transfusions, previously the only method of treatment.

To understand the significance of these advances and discoveries, it helps to understand how blood clots are usually formed. When a blood vessel is cut, it contracts to slow down the bleeding. Platelets in the blood go to the break or cut and form a clump or plug to patch the hole. The coagulation factors in the blood interact with the platelets and other chemicals in the blood to form a network or web that holds the clot in place. This complicated series of chemical reactions is called the coagulation cascade. People with hemophilia, however, have low amounts of coagulation factors. The severity of hemophilia depends on the level of the coagulation factors. A person with mild hemophilia has between 5 and 40 percent of normal coagulation factor activity; a person with moderate hemophilia has between 1 and 5 percent; a person with severe hemophilia has less than 1 percent of normal coagulation factor activity.

## **Demographics**

Hemophilia A is the more common form of the disorder, occurring in about one in every 4,000 male infants around the world. Hemophilia B affects about one in every 20,000 newborn boys. Girls who carry a defective F8 or F9 gene usually do not suffer from the disease; however, about 10 percent of girls with one abnormal copy of either defective gene will experience heavy menstrual periods and other mild problems with bleeding.

As far as is known, both hemophilia A and hemophilia B are equally common in all racial and ethnic groups around the world. About 60

## Safety of the Blood Supply

The discovery of freeze-drying techniques to separate clotting factors from whole blood in the 1960s reduced hemophiliacs' need for periodic visits to a hospital for long and costly transfusions of whole blood. Clotting factors that could be infused at home as well as in a doctor's office lengthened the life spans of hemophiliacs and also gave them more independence to lead relatively normal lives.

The situation changed abruptly in the early 1980s with the discovery of the AIDS virus. Although human blood in the United States was screened for syphilis after 1948 and hepatitis B after 1971, the blood supply had been contaminated with AIDS before screening was available. By the fall of 1982, only a few months after the Centers for Disease Control and Prevention (CDC) had issued its first bulletin about AIDS, hemophiliacs who had received clotting factors derived from human blood were being diagnosed with it. By 1985, when effective tests to screen donated blood for HIV had been developed and were in use, about 30,000 Americans, including 9,500 hemophilia patients, had already been infected via contaminated blood. Many hemophiliacs subsequently died from AIDS rather than hemophilia.

The development of genetically engineered clotting factors, made by using recombinant DNA technology without involving human blood or cells, later virtually eliminated the possibility of disease transmission. Two of the products are BeneFIX, designed to supply coagulation factor IX in patients with hemophilia B, and ReFacto, which supplies coagulation factor VIII in patients with hemophilia A. percent of persons diagnosed with hemophilia A and 44 percent of persons with hemophilia B have severe disease.

## **Causes and Symptoms**

Both hemophilia A and B are caused by genetic mutations that affect the blood's ability to clot normally. Without enough factor VIII (in the case of hemophilia A) or factor IX (in the case of hemophilia B), the platelets that move to the cut or break in a blood vessel are not held securely within a network of protein fibers. They cannot form a clot strong enough to effectively stop the bleeding, which continues for a longer period of time than in normal people.

The symptoms of hemophilia may include:

- Large or deep bruises, or unexplained bruises
- Nosebleeds that start suddenly without any obvious injury
- Tightness in the joints from blood collecting in the joint spaces
- Blood in the stools or urine
- Prolonged bleeding after minor cuts or injuries, or after routine dental work, tooth extractions, or minor surgical procedures

Patients with severe hemophilia may develop symptoms that indicate a medical emergency. These include sudden severe headache, neck pain, seeing double, repeated vomiting, or sudden pain, swelling, and warmth in the large joints (knees, elbows, hips and shoulders) or in the muscles of the arms and legs.

#### Diagnosis

The diagnosis of hemophilia depends in part on its severity. Male babies with severe hemophilia are often diagnosed shortly after birth, particularly if they are circumcised. In some cases the disorder is diagnosed when the toddler begins to walk, bruises easily, or starts having nosebleeds. Patients with milder hemophilia may not be diagnosed until they are older and have prolonged bleeding following dental work or minor surgery.

The most common test used to diagnose hemophilia is a blood test. A sample of the patient's blood is analyzed for the amount of clotting factor activity that is present. Genetic testing can also be used to diagnose people who have only mild symptoms of hemophilia A or B, as well as identify women who are carriers of hemophilia gene mutations before they become pregnant.

## Treatment

There is no cure for hemophilia. Treatment is directed at preventing severe bleeding episodes and managing symptoms when they do occur.

Patients with mild hemophilia A may be treated with injections of a hormone called desmopressin or DDAVP, which stimulates the patient's body to release more of its own clotting factor. Patients with hemophilia B or moderate to severe hemophilia A are treated with clotting factors derived from donated human blood or from genetically engineered blood products called recombinant clotting factors.

Patients with hemophilia can be taught to inject themselves with desmopressin or clotting factors at home two or three times a week as a form of prophylaxis, or preventive measure.

Patients with severe hemophilia whose joints have been damaged by bleeding usually need physical therapy to restore range of motion and strength in the damaged joints. They may eventually need to have the joints replaced with artificial joints in adult life.

## Prognosis

The life expectancy and quality of life for males with hemophilia have increased dramatically since the 1950s. Before 1960, the average life expectancy of a boy with hemophilia was 11 years. Early death was often preceded by severe pain from bleeding into the joints. As of the early 2000s, life expectancy has increased to fifty-five to sixty years. Older men with severe hemophilia who were treated in the late 1970s or early 1980s are still at risk of death from AIDS; 90 percent of these patients are HIV-positive. About 8 percent of patients with hemophilia eventually die from bleeding into the brain.

About 25 percent of children between six and eighteen years of age with severe hemophilia have below-normal academic skills and an increased risk of emotional and behavioral problems.

## Prevention

Hemophilia can be prevented in part by genetic testing of prospective parents. Although males with hemophilia cannot pass the disease on to sons, they can father daughters who will carry the disease to the next

## WORDS TO KNOW

**Coagulation cascade:** The complex process in which platelets, coagulation factors, and other chemicals in the blood interact to form a clot when a blood vessel is injured.

**Coagulation factors:** Proteins in blood plasma involved in the chain of chemical reactions leading to the formation of blood clots. They are also called clotting factors.

**Gene therapy:** An approach to treating disease by inserting healthy genes into a person's

genetic material or by inactivating defective genes.

- **Plasma:** The liquid part of blood, about 55 percent of blood by volume.
- **Platelets:** Specialized cells in the blood that are involved in forming blood clots. Platelets are also called thrombocytes.
- **Prophylaxis:** The use of a medication or other therapy to maintain health and prevent disease.

generation. Hemophilia cannot be completely eliminated by family planning, however, as 34 percent of all hemophilia A cases and 20 percent of all hemophilia B cases are caused by spontaneous mutations in each generation.

## The Future

Hemophilia will always be rare, but it is unlikely to ever be completely eliminated for two reasons. One is the role of spontaneous mutations in producing defective F8 and F9 genes in each new generation. The other reason is that most men with hemophilia now live long enough to father children and pass on the defective genes. Because of this change in life expectancy, researchers are presently concentrating on gene therapy as a possible cure for hemophilia. In gene therapy, a normal gene to replace the defective gene is inserted into the patient's genetic material by using a virus as a carrier. Research on gene therapy for hemophilia A is being conducted as of 2008.

**SEE ALSO** AIDS; Hepatitis A

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## Hepatitis A

## Definition

Hepatitis A is an infectious disease of the liver caused by the HAV virus. The disease is usually transmitted by food or water contaminated by human wastes containing the virus or by close human contact. As far as is known, only humans and some primates can get hepatitis A; it is not carried by other animals.

## Description

Hepatitis A is an inflammation of the liver caused by the HAV virus. It differs from hepatitis B and hepatitis C in that it does not cause long-term liver damage. Even though people can take several weeks or months to recover completely from hepatitis A, they have lifelong immunity afterward. Complications from hepatitis A are rare and usually limited to people with chronic liver disease or who have received a liver transplant. **Also Known As** Infectious hepatitis, viral hepatitis

#### Cause Virus

#### Symptoms

Nausea, vomiting, jaundice, fever, fatigue, abdominal pain, darkcolored urine

#### Duration

A few weeks to six to nine months



Hepatitis A varies in severity. Children and younger adults may have no symptoms at all, although they can still spread the disease. In general, adults are more likely to have noticeable symptoms than children or teenagers. The symptoms begin between two and six weeks after the person has been infected with HAV. The most common symptom is loss of energy and overall tiredness. Some people develop a mild flu-like illness with diarrhea, low-grade fever, nausea, vomiting, and muscle cramps. People with more severe symptoms may have pain in the abdomen in the area of the liver (below the rib cage on the right side of the body); they may notice that their urine has turned dark brown or that they

Magnified image of the hepatitis A virus. © PHOTOTAKE INC. / ALAMY.

have jaundice—yellowing of the skin and the whites of the eyes. Some have itchy skin.

Most people feel better within four to six weeks after the symptoms begin, although about 15 percent of patients may take up to nine months to completely regain their energy and feel normal again.

## Demographics

Hepatitis A is much more common in Africa, Asia, and South America than in the United States. The rates of hepatitis A in North America have been steadily dropping since the 1980s. In 1988 the Centers for Disease Control and Prevention (CDC) reported 32,000 cases in the United States; in 2003, 7,653 cases were reported. In developing countries, children below the age of two account for most new cases of hepatitis A; in the United States, the age group most often affected is children between the ages of five and fourteen.

Males and females are equally likely to get hepatitis A, as are people from all races and ethnic groups in the United States.

Some groups of adults are at increased risk of hepatitis A:

- People who travel to parts of the world with high rates of the disease and poor sanitation
- Male homosexuals
- People who use illicit drugs, whether injected or taken by mouth

- Medical researchers and laboratory workers who may be exposed to HAV
- Child care workers
- Homeless people

## **Causes and Symptoms**

Hepatitis A is caused by a virus that is transmitted by close personal contact with an infected person, by needle sharing, and by eating food or drinking water contaminated by fecal matter. After the virus enters the body, it multiplies in the cells of the liver, causing inflammation of the liver and a general response from the immune system that leads to most of the symptoms of the illness.

The HAV virus is shed from the liver into the bile (a digestive fluid secreted by the liver) and then into the person's stools between fifteen and forty-five days before symptoms appear. That means that people can spread the virus through their feces before they know that they are sick. In the United States, hepatitis A is most commonly spread by food handlers who do not wash their hands properly after using the bathroom; by childcare workers who do not wash their hands after changing a baby's diaper; by anal sex; and by eating raw shellfish harvested from sewage-polluted waters. In very rare cases the virus can be transmitted through blood transfusions.

In addition to fatigue, the most common symptoms of hepatitis A include:

- Low-grade fever (101°F [38.3°C])
- Nausea, vomiting, and diarrhea
- Loss of appetite and weight loss
- Swelling of the liver and pain in the area of the abdomen over the liver

## Hepatitis E

Hepatitis E is an infection of the liver caused by the hepatitis E virus, or HEV, first identified during an outbreak in New Delhi, India, in 1955, when 30,000 cases were reported following river flooding that carried raw sewage into the city's water supply. At first the disease was thought to be hepatitis A, but was later identified as a new virus. In 1990 its genetic material was analyzed and the virus was named hepatitis E.

Hepatitis E is most common in countries with tropical climates, particularly in southeastern Asia, Africa, India, and Central America. Unlike hepatitis A, which seems to affect only humans, hepatitis E has been found in deer, pigs, rats, and other animals, and can be spread by uncooked meat or shellfish. Mortality rates are generally low (around 2 percent) with hepatitis E; however, pregnant women are at high risk of liver failure from the disease. In India as many as 20 percent of pregnant women infected with hepatitis E die of liver failure.

Hepatitis E is rare in the United States, affecting primarily people who travel to countries where it is common. The disease is self-limited and is usually treated with fluid replacement; antibiotics are not effective in treating viruses. The best treatment is prevention—travelers in countries with tropical climates should avoid drinking water that has not been tested for purity and eating uncooked shellfish or raw, unpeeled fruits or vegetables.

- Tea- or coffee-colored urine
- Jaundice
- · Generalized sensation of itching
- Pale or clay-colored stools
- Muscle pains

#### Diagnosis

The doctor may suspect that a patient has hepatitis A during a physical examination by feeling the area over the liver for signs of swelling and pain; and checking the skin and eyes for signs of jaundice. A definite diagnosis is provided by a blood test for certain antibodies to the HAV virus. The doctor will also have the sample of blood checked for abnormally high levels of chemicals produced in the liver.

#### Treatment

There is no specific drug treatment for hepatitis A, as antibiotics cannot be used to treat virus infections. Most people can care for themselves at home by making sure they get plenty of fluids and adequate nutrition. People whose appetite has been affected may benefit from eating small snacks throughout the day rather than three main meals and eating soft and easily digested foods. Patients with mild vomiting may be prescribed antiemetics (drugs to control nausea). Those with severe vomiting may need to be hospitalized in order to receive intravenous fluids.

Patients with hepatitis A should avoid drinking alcohol or taking acetaminophen (Tylenol), which make it harder for the liver to recover from inflammation. Patients should also tell their doctor about any other over-the-counter or prescription drugs they are taking, because the drugs may need to be stopped temporarily or have the dosages changed.

#### Prognosis

Most people recover fully from hepatitis A within a few weeks or months. Between 3 and 20 percent have relapses (temporary recurrences of symptoms) for as long as six to nine months after infection.

About 1 percent of patients develop liver failure following HAV infection, mostly those over sixty or those with chronic liver disease. In these cases liver transplantation may be necessary for the patient's survival. There are about 100 deaths from hepatitis A reported each year in the United States.

## WORDS TO KNOW

- **Antiemetic:** A type of drug given to control nausea and vomiting.
- **Bile:** A yellow-green fluid secreted by the liver that aids in the digestion of fats.
- **Hepatitis:** The medical term for inflammation of the liver. It can be caused by toxic substances or alcohol as well as infections.

Jaundice: A yellowish discoloration of the skin and whites of the eyes caused by increased levels of bile pigments from the liver in the patient's blood.

## Prevention

Hepatitis A can be prevented by a vaccine called Havrix that is given before exposure to the HAV virus. The vaccine is given in two shots, the second given between six and eighteen months after the first. It confers immunity against hepatitis A for at least twenty years. Those who should receive the vaccine include people in the military and those who travel abroad frequently; men who have sex with other men; people who use intravenous drugs; people with hemophilia who must receive human blood products; and people who have chronic hepatitis B or C infection.

People who have been exposed to the HAV virus and children under the age of two should not be given Havrix, but they can be given another type of drug to protect them against HAV.

Everyone can reduce their risk of hepatitis A by observing the following precautions:

- Practice good personal hygiene; wash hands frequently, especially after using the toilet or changing a child's diaper.
- When traveling, drink only bottled water, avoid raw or undercooked meat or shellfish, and avoid eating fresh fruits or vegetables unless you have washed and peeled them yourself.
- Avoid sharing drinking glasses and eating utensils. If someone in the family has hepatitis A, wash their glasses and utensils separately in hot, soapy water.
- Avoid sexual contact with anyone who has hepatitis A.

## **The Future**

The rates of hepatitis A in the United States and other developed countries are likely to continue to drop, given the availability of an effective vaccine against the disease. Hepatitis A is, however, likely to continue to be a major health problem in developing countries, and travelers will need to protect themselves against it for the foreseeable future.

SEE ALSO Alcoholism; Hemophilia; Hepatitis B; Hepatitis C

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# Hepatitis B

## Definition

Hepatitis B is a viral infection of the liver transmitted through the blood or body fluids of someone who is infected. It is the most common serious liver infection worldwide. The disease has two forms: an acute form that lasts a few weeks, and a chronic form that can last for years and can lead to cirrhosis, liver failure, liver cancer, and even death. Acute hepatitis B has a 5 percent chance of leading to the chronic form of the infection in adults; however, infants infected during the mother's pregnancy have a 90 percent chance of developing chronic hepatitis B, and children have a 25–50 percent chance.

About two-thirds of people with chronic HBV infection are so-called "healthy" carriers of the virus. They may never get sick themselves but they can transmit the infection to others. The remaining one-third of people with chronic hepatitis B develop liver disease that can lead to permanent scarring of the liver. Between 15 and 25 percent of people with chronic hepatitis B eventually die of liver disease.

## Description

Hepatitis B has an incubation period of one to six months. About 50 percent of people with the acute form of the disease have no symptoms at all; the others experience loss of appetite, nausea and vomiting, and jaundice around twelve weeks after getting infected. Some patients may also have joint pain, itchy skin, or abdominal pain. Many of these patients assume that they have influenza.

Patients with chronic hepatitis may have no symptoms at all. The one-third who do eventually fall ill have the same symptoms as patients with the acute form of the disease.

People who have been infected by HBV and have recovered from the infection are protected against hepatitis B for the rest of their lives. People can also be protected by receiving a vaccine against the virus.

## Demographics

There are about 200,000 new cases of hepatitis B in the United States each year; it is estimated that 1-1.25 million people carry the disease.

Also Known As HBV infection

**Cause** Virus

#### Symptoms

Loss of appetite, jaundice, fatigue, nausea, vomiting, itchy skin; some patients have no symptoms

**Duration** Months to years



Magnified image of the hepatitis B virus. © PHOTOTAKE INC. / ALAMY.

Hepatitis B causes about 5,100 deaths in the United States each year. In the rest of the world, as many as a third of the population are chronic carriers of the disease.

The age group most commonly affected by hepatitis B in the United States is adults between the ages of twenty and fifty. African Americans are more likely to be infected than either Hispanics or Caucasians; however, Alaskan Eskimos and Pacific Islanders have higher rates of carrier status than members of other racial groups. More males than females are infected with hepatitis B in all races and age groups.

Risk factors for hepatitis B include:

- Having unprotected sex with a partner regardless of sexual orientation.
- Having a large number of sexual partners.
- Being infected with another sexually transmitted disease (STD), particularly gonorrhea or chlamydia.
- Sharing needles with other intravenous drug users.
- Having a family member with chronic HBV infection.
- Having had a blood transfusion or use of blood products before 1972.
- Needing hemodialysis for kidney disease.
- Frequent travels to parts of the world with high rates of hepatitis B. These include the Middle East, southern Africa, Southeast Asia, Brazil, and the Pacific Islands.
- Working in a hospital, clinic, or other facility requiring frequent exposure to blood, open wounds, or other body secretions.

## **Causes and Symptoms**

Hepatitis B is caused by a virus. It is primarily a bloodborne infection, but can also be transmitted through contact with the semen or saliva of an infected person. The virus enters the body through injection, a break in the skin, or contact with the mucous membranes, tissues that line the mouth, genitals, and rectum. People cannot get hepatitis B from food or from shaking hands, sneezing or coughing, breastfeeding, or casual contact with an infected person.

The symptoms of acute hepatitis B infection include:

- Loss of appetite
- Feeling tired
- Muscle and joint aches
- Low-grade fever
- Abdominal pain in the area below the rib cage
- Yellowish discoloration of the skin and whites of the eyes
- Tea- or cola-colored urine
- Grayish or clay-colored stools

A few people develop a severe form of hepatitis B known as fulminant hepatitis. This form of the disease appears rapidly and can cause death. Its symptoms include:

- Sudden collapse
- Mental confusion, hallucinations, or extreme sleepiness
- Jaundice
- Noticeable swelling of the abdomen

## Hepatitis D

Hepatitis D, or delta hepatitis, is a liver disease caused by a virus (HDV) unrelated to the viruses that cause hepatitis A, B, and C. HDV is sometimes called the delta agent. Discovered only in 1977, the hepatitis D virus can replicate (multiply) only in patients infected with HBV. As of 2008 HDV infection was found in about 5 percent of patients diagnosed with hepatitis B. Hepatitis D can range from an acute, but limited, infection to a sudden, severe disease that leads to liver failure in 1 percent of patients. Most patients who are infected with hepatitis D alongside hepatitis B eventually get better. Chronic infection with both viruses occurs in only 5 percent of patients.

The World Health Organization (WHO) estimates that there are about fifteen million people worldwide infected with HDV. In the United States, hepatitis D is primarily a disease of adults rather than children. Risk factors include intravenous drug use and coming from southern Italy or other countries around the Mediterranean Sea.

HDV infection produces the same symptoms as hepatitis B. It can be detected by a blood test known as the anti-delta agent antibody test. Treatment is the same as for hepatitis B.

## Diagnosis

Hepatitis B is diagnosed by one or more blood

tests, since patients may not have any apparent symptoms. In a number of cases, the person is diagnosed following a routine blood test given as part of an annual health checkup. The most common clue is abnormal liver function results.

To confirm the diagnosis, the doctor will take one or more blood samples for testing:

- A test of liver function, if this has not already been done.
- Tests for antibodies to the hepatitis B virus. A positive result means that the person has either been effectively vaccinated against

HBV or has been infected at some point in the past and has recovered.

- Tests for the surface antigen of the hepatitis B virus (HBsAg). The surface antigen is the outer coating of the virus. A positive HBsAg test means that the patient is currently infected and may be able to pass on the virus to others.
- Hepatitis B DNA test. This blood test measures the levels of virus in the patient's blood.

Patients with chronic active hepatitis B may be given a computed tomography (CT) scan or ultrasound of the liver to see whether the liver has been damaged by the infection. The doctor may also perform a liver biopsy. This test involves inserting a long hollow needle into the patient's liver through the abdomen and withdrawing a small amount of tissue for examination under a microscope.

## Treatment

Patients who know that they have been exposed to the hepatitis B virus can be treated by administering an immune-boosting injection and three shots of the HBV vaccine to prevent them from developing an active infection. Those who have already developed symptoms of the acute form of the disease may be given intravenous fluids to prevent dehydration or anti-nausea medications to stop vomiting. To date, there is no medication that can prevent acute hepatitis B from becoming chronic once the symptoms begin.

There are few treatment options for chronic hepatitis B. If the patient has no symptoms and little sign of liver damage, the doctor may suggest monitoring the levels of HBV in the patient's blood periodically rather than starting treatment right away. There are five different drugs used to treat hepatitis B, but they do not work in all patients and may produce severe side effects. Most doctors will wait until the patient's liver function begins to worsen before administering these drugs.

If the patient develops fulminant hepatitis B or their liver is otherwise severely damaged by HBV, the only option is a liver transplant. This is a serious operation with a lengthy recovery period; its success also depends on finding a suitable donor liver.

## Prognosis

Patients with acute hepatitis B usually recover; the symptoms go away in two to three weeks, and the liver itself returns to normal in about four months. Other patients have a longer period of illness with very slow improvement. Chronic hepatitis leads to an increased risk of cirrhosis and liver cancer, and eventual death in about 1 percent of cases.

## Prevention

Hepatitis B can be prevented by vaccination with a vaccine called Engerix-B. The person receives the first two doses of the vaccine a month apart and the third dose six months later. The vaccine is recommended for all persons under the age of twenty; it can be given to newborns and infants as part of their regular vaccination series. Others who should be vaccinated include health care workers, military personnel, firefighters and police, people who travel frequently to countries with high rates of hepatitis B, people with hemophilia, people who must be treated for kidney disease, people who inject illegal drugs, and men who have sex with other men.

Other preventive measures include:

- · Practicing safe sex
- Not sharing needles, razors, toothbrushes, or any other personal item that might have blood on it
- Avoiding getting a tattoo or body piercing, as some people who perform these procedures do not sterilize their needles and other equipment properly
- Getting tested for HBV infection if pregnant, as the virus can be transmitted from a mother to her unborn baby
- Consulting a doctor before taking an extended trip to any country with high rates of hepatitis B

## The Future

The rate of hepatitis B in the United States began to drop after 1992, when vaccination of infants became routine, followed by vaccination of adolescents in 1995. Public health doctors expect the decline to continue for the foreseeable future.

Researchers at the National Institutes of Health (NIH) are presently looking for medications that will be effective in treating all patients with chronic hepatitis B with fewer side effects.

SEE ALSO Hemophilia; Hepatitis A; Hepatitis C

## WORDS TO KNOW

- **Carrier:** A person who is infected with a virus or other disease organism but does not develop the symptoms of the disease.
- Chronic: Long-term or recurrent.
- **Cirrhosis:** Disruption of normal liver function by the formation of scar tissue and nodules in the liver.
- **Fulminant:** Referring to a disease that comes on suddenly with great severity.

- **Hepatitis:** A general term for inflammation of the liver. It can be caused by toxic substances or alcohol as well as infections.
- Jaundice: A yellowish discoloration of the skin and whites of the eyes caused by increased levels of bile pigments from the liver in the patient's blood.

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# Hepatitis C

## Definition

Hepatitis C infection is an inflammatory disease of the liver caused by HCV. HCV is most commonly transmitted from person to person through contaminated blood.

## Description

Hepatitis C is an infection that often goes undetected until it has done significant damage to a patient's liver. The infection has two phases, acute (the first six months) and chronic (after the first six months). A minority of patients clear the virus from their bodies during the acute phase, but 60–85 percent have a chronic hepatitis C infection.

People may have no symptoms of illness during the acute phase of hepatitis C infection and possibly only a mild flu-like syndrome later. Symptoms of severe liver damage, such as nausea, vomiting, collection of fluid in the abdomen, and mental changes, may not develop for ten or twenty years after the initial infection.

## Demographics

Hepatitis C is the major source of chronic liver infection in North America. There are approximately 30,000 new infections and 8,000–10,000 deaths each year in the United States. It is estimated that 4 million persons in the United States have been infected by HCV and 2.7 million of these have the chronic form. HCV infection presently accounts for 40 percent of referrals to liver clinics.

HCV is more common among Hispanics and African Americans than among Caucasians, Asian Americans, or Native Americans. Sixtyfive percent of persons with HCV infection are between thirty and forty-nine years of age.

People who are at increased risk of HCV include:

- Those who abuse intravenous drugs (60 percent of new cases)
- · People who have unprotected sex with a large number of partners
- · People who require hemodialysis for kidney disorders
- People who need frequent blood transfusions

Also Known As Non-A, non-B hepatitis

Cause Bloodborne virus

#### Symptoms

Fatigue, nausea, vomiting, jaundice, dark urine, liver pain, joint and muscle pains

#### Duration

May be lifelong after infection



Magnified image of the hepatitis C virus. © PHOTOTAKE INC. / ALAMY

- People who are HIV-positive
- Health care workers who may get needlestick injuries

## **Causes and Symptoms**

Hepatitis C is caused by HCV. It is most often transmitted from one person to another through infected blood or blood products, but can also be (uncommonly) transmitted from mother to child during childbirth or through sexual intercourse. Before 1992, HCV was sometimes transmitted through blood transfusions, hemodialysis, or transplanted organs from infected donors; these are now rare events. In 1992, researchers invented a new test for checking blood products for HCV; as a result, new infections annually in the United States declined from 240,000 in the 1980s to about 20,000–30,000 in 2007. The most common cause of HCV transmission is intrave-

nous drug use; transfusion-related cases of hepatitis C now occur only once in every 2 million transfused units of blood.

Hepatitis C infection is sometimes divided into an early phase called the acute stage and a later phase called the chronic stage. The acute stage begins when the virus enters the body; it lasts for about six months. Antibodies to the virus can usually be detected between three and twelve weeks after infection. About 15–40 percent of people who are infected clear the virus from their bodies during this phase, while the other 60–85 percent go on to develop chronic hepatitis C infection. It is this second group of patients who run the risk of suffering cirrhosis or other forms of liver or kidney damage years later.

Eighty percent of patients infected by HCV in its early stage do not have any symptoms, or have mild and nonspecific symptoms like fatigue. Others have a flu-like syndrome marked by poor appetite or nausea, soreness in the area of the liver, or pains in the joints and muscles. Some may notice that their urine is dark and looks like tea or cola. If chronic HCV infection leads to liver disease ten to twenty years later, the patient may have the following symptoms:

- Severe loss of appetite
- Nausea and vomiting, with blood in the vomit

- Low-grade fever
- Itchy skin
- Jaundice (This is a yellowish discoloration of the whites of the eyes and the skin caused by an increase in the amount of bile pigments from the liver in the patient's blood.)
- Sleep disturbances
- Swelling of the abdomen caused by fluid retention
- Diarrhea
- Difficulty urinating
- Confusion, hallucinations, difficulty concentrating, or other mental disturbances

## Diagnosis

Diagnosis of hepatitis C infection is often delayed for years because many patients with chronic hepatitis C infection do not have noticeable or troublesome symptoms until liver damage has already occurred. In some cases a person with chronic hepatitis C infection is detected through routine blood testing for abnormal liver function or because they have a history of intravenous drug abuse or HIV infection. Testing for chronic infection begins with blood tests that indicate the presence of antibodies to HCV. Since antibody tests cannot tell whether the person is currently infected, however, a second blood test that looks for the virus's characteristic genetic material is performed.

An Outbreak in Nebraska

Transmission of hepatitis C virus (HCV) within health care facilities is never supposed to occur. In 2000–2001, however, an outbreak in Nebraska, traced to poor health safeguards at a private cancer clinic, affected almost 100 patients. A physician's wife who began treatment for breast cancer at the clinic in 2001 was surprised to find out in 2002 that she was infected with HCV. Her husband discovered on checking his records that several of his patients were also infected; all were undergoing cancer therapy at the same clinic.

An investigation by the Nebraska Health and Human Services System found that in March 2000 a clinic nurse used a syringe to rinse a known HCV patient's chemotherapy port with saline solution then used the same syringe to draw more saline from a large common container, thus transferring HCV from the infected port into the container. The virus was transmitted to other patients via the saline solution and repeated use of contaminated syringes.

Between March 2000 and June 2001, when the nurse left the clinic, 99 of the clinic's 857 patients were infected with HCV. As of 2008, at least one had died. Disturbingly, the Nebraska case is not alone; the Centers for Disease Control and Prevention (CDC) recorded at least 32 outbreaks of HCV in the United States since 1999. In March–May of 2008, 84 patients contracted HCV at an endoscopy center in Las Vegas, Nevada—again as the result of reusing contaminated syringes.

If the results are positive for both tests, the doctor will order a third blood test that determines the virus's specific genotype or genetic makeup. There are six known genotypes of HCV as of 2008, and knowing which type is involved helps to guide the patient's treatment. To determine the extent of damage to the patient's liver, the doctor may order a liver biopsy. In this procedure, a needle is inserted into the patient's liver through the abdomen in order to remove a small sample of tissue for analysis.

#### Treatment

Not all patients with HCV require therapy, but if treatment is needed, the first line of treatment comprises two medications known as Interferon, a drug that resembles some of the proteins that the body makes naturally to fight viruses, and Virazole, which is an antiviral drug. The combination of these drugs works better than Interferon alone. Interferon is usually given as a shot once a week and Ribavirin is taken as a pill twice a day. The length of treatment depends on the genotype of HCV; patients with genotype 2 or 3 are treated for twenty-four weeks whereas patients with genotype 1 or 4 must undergo forty-eight weeks of treatment. The cure rates for genotypes 1 and 3 are about 75 percent; the cure rate for genotype 1 is 50 percent; and for genotype 4 it is 65 percent. Unfortunately, Interferon and Ribavirin produce unpleasant side effects for patients that range from depression and irritability to weight loss, nausea, and muscle pains. In addition to side effects, Ribavirin cannot be given at all to pregnant women because it can harm the unborn child.

The only treatment for cirrhosis or severe liver disease is liver transplantation. The problem, however, is that there are many more patients waiting for donated livers than there are suitable organs available. In addition, liver transplantation does not cure hepatitis C infection; most people who receive transplanted livers will develop a recurrence of the virus. The effectiveness of medication treatment of hepatitis C following a liver transplant is unclear.

Patients with chronic hepatitis C should stop drinking alcohol, as it can speed up the rate of liver damage. They should also be vaccinated against hepatitis A and hepatitis B.

#### Prognosis

According to the CDC, between 75 and 85 percent of people infected with HCV will develop chronic hepatitis C infection. Twenty percent of these chronically infected persons will develop cirrhosis of the liver within twenty years of infection; 1–5 percent of chronically infected people will eventually die of liver disease.

## WORDS TO KNOW

Chronic: Recurrent or long-term.

**Cirrhosis:** Liver damage most commonly caused by alcoholism or hepatitis C.

**Genotype:** The genetic makeup of a cell or organism.

- **Hepatitis:** Inflammation of the liver. It can be caused by toxic substances or alcohol as well as infections.
- Jaundice: A yellowish discoloration of the skin and whites of the eyes caused by increased levels of bile pigments from the liver in the patient's blood.

Women with chronic hepatitis C have better outcomes than men, and patients infected at younger ages have better outcomes than those infected in middle age. The reason for these differences is not clear.

## Prevention

There is no vaccine that can prevent HCV. Prevention depends on careful observation of good health practices in hospitals and clinics and on individual lifestyle changes. The CDC recommends the following ways that individuals can lower their risk of getting hepatitis C:

- Do not use intravenous drugs. People who cannot quit should never share their needles, syringes, water, or other materials used to inject drugs. They should also get vaccinated against hepatitis A and hepatitis B.
- Do not share personal items (razors, toothbrushes, etc.) that might have blood on them.
- Avoid getting tattoos or body piercing. People who do get a tattoo, however, should at least make sure that the operator who performs the tattoo is using proper sterile procedure.
- Use latex condoms when having sex. Although it is rare for HCV to be transmitted through sexual intercourse, it can happen.
- People who discover that they are infected with HCV should not donate blood, organs, or tissues.

## The Future

Researchers expect the number of people who die from hepatitis C in the United States to increase in the following years. One researcher found that the number of deaths due to HCV-related complications rose from fewer than 10,000 in 1992 to almost 15,000 in 1999. This number is expected to rise in the years ahead because of the growing numbers of people with HIV and other chronic infections. These infections increase a person's risk of developing severe liver disease if they do become infected with HCV.

Researchers are continuing to work on an effective vaccine against HCV and more effective medications with fewer side effects for treating the chronic form of the disease.

#### SEE ALSO Hepatitis A; Hepatitis B

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## High Cholesterol See Hypercholesterolemia.

**HIV Infection** 

See AIDS.



## **Hives**

## Definition

Hives refers to an eruption of wheals (flat-topped itching or stinging bumps, welts, or patches) on the skin following contact with an allergen or some other physical agent. It is not a single disease but rather a pattern of reaction to an irritant of some kind. Hives may be caused by infections, medications, insect bites, chemicals, food allergies, underlying medical disorders, or a variety of other causes. Hives that last less than six weeks are defined as acute urticaria; hives that are present for six weeks or longer or that recur frequently are known as chronic urticaria.

## Description

Hives are pale, itchy, or stinging welts or wheals that form on the skin, most commonly as a reaction to an irritant. Hives located near the mouth, eyes, or genitals may cause the lower layers of nearby skin to swell up or look puffy—this reaction is known as angioedema.

Hives vary in size from about a quarter-inch in diameter to six inches or even larger. They can also join together to form even larger patches of Also Known As Urticaria, nettle rash

#### Cause

Allergy, exposure to cold or scratching, reaction to medication

#### Symptoms

Itching, burning, or stinging patches or wheals on the skin

#### Duration

A few hours to years, depending on type



*Hives on a patient's back, a result of a strep infection.* © PHIL DEGGINGER / ALAMY.

raised skin with clearly defined edges. They will turn white when the center of the wheal is pressed. They may appear suddenly, change shape, and disappear within hours or even minutes. Hives, after disappearing, do not leave scars on the skin.

Acute hives are typically caused by an allergic reaction to foods or medications, fresh foods being more likely to cause hives than cooked foods. The most common "problem" foods are nuts, chocolate, fish and shellfish, tomatoes, eggs, fresh berries, soy, wheat, and milk. Almost any medication can cause hives in a susceptible person. The common offenders are aspirin and ibuprofen; penicillin and other antibiotics; codeine and pain relievers containing codeine; and medications given to treat high blood pressure. Other triggers of acute hives include certain substances, particularly latex; pollen, molds, and animal dander; infections, particularly the common cold; and insect bites.

Some cases of acute hives are triggered by physical causes, such as exposure to sunlight, hot weather, cold water, or exercise. Hives resulting from sun exposure usually fade in one to two hours. Some people who get hives after swimming in cold water may feel faint or dizzy as well. A few people get hives when their skin is scratched or stroked firmly. This condition is called dermatographism and can occur together with hives caused by allergens.

Chronic hives last for longer than six weeks. The causes of chronic urticaria are usually more difficult to identify than the causes of acute

hives. They often include autoimmune disorders like lupus or Hashimoto disease, or bacterial or fungal infections. In the majority of cases, however, the cause of chronic hives is never discovered. Chronic hives may last for months or even years and have a severe impact on the patient's quality of life.

## **Demographics**

Hives are a common skin problem in North America, affecting between 20 and 25 percent of people at some point in their lives. As far as is known, hives are equally frequent in all races and ethnic groups, although they are more common in women—there are three females for every two males affected. Chronic urticaria is less common that acute urticaria, affecting between 1 and 3 percent of the population. Dermatographism affects about 5 percent of the general American population.

Acute hives can develop in people of any age, but are most common in teenagers and young adults in their twenties. Chronic hives, on the other hand, are more common in middle-aged adults.

## **Causes and Symptoms**

The basic cause of hives is the release of a chemical called histamine. The histamine causes small blood vessels to leak plasma (the liquid part of blood) into the skin, causing the surface of the skin to rise and form wheals or angioedema.

The symptoms of hives are itching, stinging, and burning on the affected parts of the skin. In some cases, angioedema can affect the tissues

## **Home Treatment for Hives**

While waiting for antihistamine and other medications to take effect, patients with hives can do the following to relieve itching and speed the healing of their skin:

- Avoid taking hot baths or showers; use lukewarm water instead.
- Avoid using harsh or heavily perfumed soaps.
- Stay out of direct sunlight during the hottest hours of the day; exercise during the early morning or early evening.
- Avoid heavy exercise in hot weather or any activity that might cause sweating, as perspiration can make hives worse.
- Wear light, loose-fitting clothing.
- Apply cool compresses to the affected area.
- Try to work or sleep in a cool room.
- Lower the stress level if possible.

If any of the following symptoms appear with hives, a doctor should be contacted *at once* or the patient taken to an emergency room:

- Dizziness or fainting
- Difficulty in breathing or swallowing
- Wheezing
- Tightness or pain in the chest
- Swelling of the tongue, lips, or entire face
- Nausea, vomiting, abdominal cramps, or diarrhea

that line the throat, causing the airway to swell shut and make it difficult to breathe or swallow. This type of angioedema is a medical emergency and requires immediate treatment in an emergency room.

## Diagnosis

Diagnosis of the cause of acute hives is usually helped by asking patients about their food or medication history, recent exposure to sun or insect bites, recent infections, or their occupation. Patients may be asked to keep a record of the foods they eat, the medications they use, and the types of exercise they do on a regular basis. Doctors may also ask patients to keep a record of the location of the hives when they appear and how long the individual hives last.

Although it is usually not necessary to test for the causes of acute hives in order to treat the problem, the doctor may order skin tests to see whether specific foods, medications, or chemicals are triggering the hives. In some cases the hives may have more than one cause. Blood tests may be ordered to test for the possible causes of chronic hives. A primary care doctor may refer the patient to a dermatologist (a specialist in skin diseases) for more detailed tests or to make sure that the patient does not have a different type of skin disease.

## Treatment

Acute hives can often be treated at home by a combination of antihistamine medications prescribed by the doctor, avoiding known triggers, and protecting the skin against further irritation (see sidebar). If the hives are not relieved by antihistamines, the doctor may prescribe cortisone or other steroid medications, but these should be used only for short periods of time.

Patients who have severe attacks of hives or angioedema are usually given an injection of adrenaline to clear the airway and are taken to a hospital emergency room. People who have repeated episodes of angioedema that cause breathing problems are usually given an EpiPen, which is a device that contains adrenaline, so that they can inject themselves in an emergency.

Patients whose hives appear to be triggered by emotional stress may be helped by relaxation techniques or stress management programs.

## Prognosis

The prognosis of hives depends on the cause and whether the hives are acute or chronic. Acute hives often clear up rapidly once the allergen or other cause of the skin reaction has been removed. Chronic hives, however, can be difficult to treat or cope with, particularly if the skin eruptions are related to an autoimmune disorder. One study found that

## WORDS TO KNOW

- **Adrenaline:** A hormone that can be used in medicine to open the breathing passages in patients with severe angioedema. It is also called epinephrine.
- Allergen: Any substance that can provoke an allergic reaction in susceptible individuals.
- **Angioedema:** The medical term for the swelling of tissues around the eyes, lips, and genitals that sometimes accompanies hives.

Chronic: Long-term or recurrent.

- **Dander:** Tiny skin, feather, or fur particles from household pets that cause allergic reactions in some people.
- **Dermatographism:** A type of hives produced by scratching or stroking the skin.
- **Dermatologist:** A doctor who specializes in diagnosing and treating diseases of the skin.
- **Histamine:** A compound that is released during an allergic reaction.
- Urticaria: The medical term for hives.
- Wheal: A suddenly formed flat-topped swelling of the skin; a welt.

chronic hives lasted a year or longer in more than 50 percent of patients and 20 years or more in 20 percent of them.

## Prevention

There is no known way to prevent all possible types of hives, although avoiding foods and medications known to trigger acute hives can help. The limitation of this approach, however, is that specific triggers cannot be identified in about 50 percent of cases.

Another recommendation is to cut down on alcoholic beverages they can make the itching of hives worse or even trigger hives in some people. Some patients are also advised by their doctors to take antihistamines on a daily basis in order to prevent acute attacks of hives.

## The Future

Both acute and chronic hives are likely to be common skin problems for the foreseeable future. It may be that further research will shed light on the causes of chronic hives so that more effective treatments can be developed.

SEE ALSO Allergies; Anaphylaxis; Hashimoto disease; Lupus

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# Hodgkin Disease

## Definition

Hodgkin disease, also known as Hodgkin lymphoma or HL, is a cancer of the blood and lymphatic system. It is the most common type of blood cancer and the third most common childhood cancer. The disease is named for Thomas Hodgkin (1798–1866), the British doctor who first described it in 1832.

## Description

Hodgkin disease is one of the two major types of lymphoma, a form of cancer that originates in lymphocytes, which are a specific type of white blood cells in the immune system. It is unusual in that it primarily affects two different age groups, adolescents and young adults between the ages of fifteen and thirty-five, and older adults over age fifty-five. Hodgkin begins in the lymphatic system, a group of organs and tissues that are part of the immune system and also help to form new blood cells. The lymphatic system includes lymph nodes, small organs composed of

## Also Known As

Hodgkin lymphoma, HL

#### Cause Unknown

#### Symptoms

Swollen lymph nodes, night sweats, fever, weight loss, lack of energy

#### Duration

Lifelong unless treated

#### Hodgkin Disease

*Image of cancer cells dividing in Hodgkin disease.* © DR. ANDREJS LIEPINS/SPL/ PHOTO RESEARCHERS, INC.



lymphoid tissue located at various points throughout the body that are joined by lymphatic vessels; the spleen, a small organ on the left side of the abdomen; the bone marrow; and the thymus gland, which is just below the neck.

Hodgkin disease begins when a type of lymphocyte called a B cell turns into an abnormal form called a Reed-Sternberg cell. These abnormal cells continue to reproduce themselves until they form a tumor. In most cases this first tumor develops in one of the lymph nodes above the diaphragm (the thin sheet of muscle that divides the chest cavity from the abdominal cavity); however, the tumor can develop in a group of lymph nodes or elsewhere in the lymphatic system. The disease usually spreads from lymph node to lymph node along the lymphatic vessels rather than skipping around the body. In the later stages of the disease, it spreads outside the lymphatic system to other organs.

There are two basic subtypes of Hodgkin disease: classic, which accounts for 95 percent of cases, and nodular lymphocyte predominant Hodgkin disease (NLPHD), which accounts for the remaining 5 percent. NLPHD differs from classic Hodgkin diseae in that it occurs primarily in lymph nodes in the neck and armpits, and its large abnormal cells are known as "popcorn" cells because they look like pieces of popped popcorn under the microscope.

## **Demographics**

Hodgkin lymphoma is a relatively rare form of cancer, affecting two or three people in every 100,000. It accounts for less than 1 percent of cancers worldwide. As of 2008, there were about 500,000 people in the United States with some form of lymphoma, but only 143,000 had Hodgkin disease. According to the National Cancer Institute (NCI), about 8,200 new cases of HL are diagnosed in the United States each year, and 1,350 people die from the disease annually.

The rates for Hodgkin disease vary according to race and sex. The highest rates are in Caucasian males (3.2 cases per 100,000), followed by African American males (3.0 cases per 100,000), Caucasian females (2.6 cases per 100,000), African American females (2.1 cases per 100,000), Asian American males (1.4 cases per 100,000), and Asian American females (1 case per 100,000). About 10 percent of cases of Hodgkin are diagnosed in children below the age of fourteen; 85 percent of these are boys.

Although the cause of Hodgkin disease is still unknown, researchers have identified several factors that increase a person's risk of developing this form of cancer:

- Age between fifteen and thirty-five or over fifty-five
- Male sex
- Infection with Epstein-Barr virus or infectious mononucleosis
- Family history of Hodgkin disease
- HIV infection
- Organ transplantation or other reasons for therapy that suppresses the immune system

#### **Causes and Symptoms**

Although researchers know that Hodgkin disease begins with the formation and multiplication of abnormal cells, they do not yet know what triggers this formation. Some scientists think that certain types of viral infections, genetic factors, or environmental toxins might be involved.

The symptoms of Hodgkin disease include:

- Swelling of the lymph nodes in the neck, armpits, or groin. These are usually painless.
- Tiredness that does not go away.
- Fever and chills.

- Night sweats.
- Unexplained weight loss of 10 percent or more of body weight.
- Itchy skin.
- Loss of appetite.

Some patients also experience heavy sweating, coughing or chest pain, difficulty breathing, enlargement of the spleen, hair loss, or neck pain.

## Diagnosis

The doctor usually begins by taking a medical history, because there are many causes other than Hodgkin for swollen lymph nodes, fever, and some of the other early symptoms of the disease. In some cases, a patient with classic Hodgkin has no early symptoms, and the disease is discovered during a routine chest x ray.

In most cases the disease is diagnosed by a tissue biopsy. The doctor removes a small piece of tissue from a swollen lymph node either by cutting directly into the swollen node or by withdrawing a tissue sample through a fine needle. In addition to looking for abnormal cells in the tissue sample, the doctor may use certain types of chemical tests to look for proteins attached to the surface of the abnormal cells.

In addition to a tissue biopsy, the doctor may order blood tests or a bone marrow biopsy. Blood tests are useful in evaluating the type of chemotherapy that would be best for the patient. In addition, imaging tests can be used to determine the location and extent of the disease. These tests may include x-ray studies, magnetic resonance imaging (MRI), computed tomography (CT) scans, positron emission tomography (PET) scans, and a gallium scan. This last type of test uses a radioactive element given intravenously to identify affected lymph nodes.

## Treatment

The first step in treating any kind of cancer is called staging. Staging is a description of the location of the cancer, its size, how far it has penetrated into healthy tissue, and whether it has spread to other parts of the body. Hodgkin disease is classified into four stages:

- Stage I: The disease is limited to one lymph node.
- Stage II. The disease involves two or more lymph nodes on the same side of the diaphragm.

- Stage III. The disease has spread to lymph nodes on both sides of the diaphragm and may involve the spleen.
- Stage IV. The disease has spread to one or more organs outside the lymphatic system, such as the lungs or liver.

The next steps in treatment depend on the stage of the disease.

- Stage I and Stage II Hodgkin disease can be treated with radiation therapy, chemotherapy, or a combination of both. Only low-dose radiation therapy is used in girls and women, because standard doses of radiation will increase their risk of breast cancer in later life.
- Stages III and IV. Chemotherapy may be used by itself to treat advanced-stage Hodgkin as well as being combined with radiation therapy. There are five different combinations of drugs that are currently used; some of these are quite toxic and have severe side effects. The drug combination preferred by most doctors in the United States is called ABVD, the initials of the four drugs that are administered.

Patients who are not cured by radiation therapy and chemotherapy, or whose disease recurs after chemotherapy, may be treated with a combination of high-dose chemotherapy and bone marrow transplantation. The bone marrow transplantation is needed because the high doses of anti-cancer chemicals will damage the patient's own bone marrow and lead to a life-threatening shortage of the red and white blood cells produced in the bone marrow.

## Prognosis

Hodgkin disease is potentially curable, although the treatments that are currently used increase the patient's risk of developing a second type of cancer later in life. As of 2008, 90 percent of patients with Stage I or II disease lived for five years after treatment. The five-year survival rates for Stages III and IV are 84 percent and 65 percent respectively. Between 20 and 25 percent of all patients diagnosed with Hodgkin will die of the disease. In the first fifteen years after treatment, recurrent Hodgkin is the major cause of death among patients; after fifteen years, death from other causes is more common.

## Prevention

There is no way to prevent Hodgkin disease, because its causes are still unknown.

## WORDS TO KNOW

- **Diaphragm:** A sheet of muscle extending across the bottom of the rib cage that separates the chest from the abdomen.
- **Lymph nodes:** Small rounded masses of lymphoid tissue found at various points along the lymphatic vessels.
- **Lymphocyte:** A type of white blood cell that fights infection. Lymphocytes are divided into two types, T cells (produced in the thymus gland) and B cells (produced in the bone marrow).
- **Popcorn cell:** An abnormal cell found in nodular lymphocyte predominant Hodgkin disease (NLPHD).
- **Reed-Sternberg cell:** An abnormal type of B lymphocyte that is found in classic Hodgkin lymphoma.
- **Staging:** Measuring the severity or spread of a cancer.
- **Thymus:** A small organ located behind the breastbone that is part of the lymphatic system and produces T cells.

## **The Future**

Current research on Hodgkin disease is focused on the possible role of viruses in causing the disease and on exploring newer forms of treatment. Although radiation therapy and chemotherapy are highly effective treatments for this form of cancer, they have a number of long-term as well as short-term side effects. Because Hodgkin disease is a relatively rare form of cancer and because there are still unanswered questions about the best way to treat it, researchers are actively looking for Hodgkin patients to participate in clinical studies.

SEE ALSO Infectious mononucleosis; Lymphoma

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## **HPV** Infection

## Definition

HPV infection is a sexually transmitted disease (STD) caused by thirty to forty of the 130 or so known strains of human papillomavirus, the name of a group of viruses that infect the skin and mucous membranes of humans and some animals. In humans these sexually transmitted strains can cause genital warts, precancerous changes in the tissues of the female vagina, or cervical cancer. Other strains of HPV are responsible for warts on the soles of the feet (plantar warts), common warts on the hands, and flat warts on the face or legs.

## Also Known As

Human papillomavirus infection, genital warts

**Cause** Human papillomavirus

#### Symptoms

None in most people; genital warts, precancerous tissue changes, cervical cancer

#### Duration

Months or years after infection

#### Description

HPV infection is one of the most common sexually transmitted diseases in the United States. Most people who are infected with one of the sexually transmitted strains of the virus, however, do not know that they have it because they have no symptoms. They can easily transmit the infection to a partner.

The various strains of HPV are classified as either low-risk or highrisk according to their potential for causing cancer or other serious health



Image of cervical cells infected with HPV. The nucleus of the cell is the black dot; in HPV infection, some cells have more than one nucleus. DR. E. WALKER / PHOTO RESEARCHERS, INC.

problems. Low-risk strains of HPV may produce genital warts but do not cause cancer. Low-risk types 6 and 11 are responsible for 90 percent of cases of genital warts. High-risk strains of the virus, which include types 16, 18, 31, and 45, may cause cervical cancer in women as well as cancers of the anus and penis in men. These high-risk strains can also cause cancer of the mouth and throat in people who have oral sex. HPV types 16 and 18 are found in more than 70 percent of cervical cancers in women.

## **Demographics**

In recent years HPV infection has become one of the most common STDs in the United States. Approximately 20 million Americans were infected with HPV as of 2008, and another 6.2 million people become newly infected each year. According to one study, 27 percent of women between the ages of fourteen and fifty-nine are infected with one or more types of HPV. The Centers for Disease Control and Prevention (CDC) estimates that more than 80 percent of American women will contract at least one strain of genital HPV by age fifty. About 75–80 percent of sexually active Americans of either sex will be infected with HPV at some point in their lifetime.

As far as is known, men and women are at equal risk of being infected with HPV, as are members of all races and ethnic groups.
Some people are at greater risk of sexually transmitted HPV than others:

- Gay and bisexual men.
- People with HIV or other diseases that weaken the immune system.
- Males or females below age twenty-five. Younger people appear to be more biologically vulnerable to the HPV virus.
- People who have large numbers of sexual partners.
- People in relationships with partners who have sex with many other people.

In terms of specific illnesses associated with HPV, 11,000 women are diagnosed with cervical cancer each year in the United States and 3,900 women die annually of the disease. Another 5,800 women are diagnosed with cancers of the vagina and the external female genitals, while 3,300 men are diagnosed with cancer of the penis or the anal area. The risk of anal cancer is seventeen to thirty-one times higher among gay and bisexual men than among heterosexual men.

#### **Causes and Symptoms**

The cause of sexually transmitted HPV infection is one or more strains of the human papillomavirus. The virus enters the body through small breaks in the skin surface or in the mucous membranes lining the genitals. In most cases the body fights off the virus within a few weeks. In some people, however, HPV remains dormant for a period ranging from a few weeks to several years in one of the lower layers of skin cells. The virus then begins to replicate (copy itself) when these cells mature and move upward to the surface of the skin. The virus affects the shape of the cells, leading to the formation of noticeable warts, precancerous changes in skin cells, or cervical cancer. About 1 percent of sexually active adults in the United States have genital warts at any one time; about 10 percent of women with high-risk HPV in the tissues of their cervix will develop long-lasting HPV infections that put them at risk for cervical cancer.

Symptoms of sexually transmitted HPV infection may include:

• Genital warts. These appear as bumps or clusters of fleshy outgrowths around the anus or on the genitals. Some may grow into larger cauliflower-shaped masses. Genital warts usually appear within weeks or months after sexual contact with an infected person. If left untreated, genital warts may go away, remain unchanged, or increase in size or number but will not turn into cancers. It is possible, however, for a person to be infected with a high-risk strain of HPV as well as one of the strains that cause genital warts; therefore the appearance of genital warts does not necessarily mean that the person is not at risk of cancer.

- Precancerous changes in the tissues of the female cervix. These are flat growths on the cervix that cannot be seen or felt by the infected woman.
- Cancer. High-risk strains of HPV can cause cancers of the mouth and throat as well as cancers of the anal area and the male and female genitals. These typically take years to develop after infection. In men, symptoms of anal cancer may include bleeding, pain, or a discharge from the anus, or changes in bowel habits. Early signs of cancer of the penis may include thickening of the skin, tissue growths, or sores.

#### Diagnosis

There is no general blood, urine, or imaging test for HPV infection. The diagnosis of genital warts is obvious based on their location and appearance. The doctor may, however, use a vinegar solution to identify HPV-infected areas on the skin of the genitals. The vinegar solution may turn white if HPV is present. Since genital warts are caused by low-risk strains of HPV, the doctor does not need to identify the specific strain of the virus that is present.

Sexually active women should be screened periodically for the presence of changes in the tissues of the cervix. The most common test is the Papanikolaou test or Pap smear, invented by a Greek physician in the 1940s. To perform a Pap smear, the doctor takes a small spatula to obtain cells from the outer surface of the cervix and smears the collected cells on a slide that is then examined in a laboratory for signs of any abnormal cells. If abnormal or questionable cells are found, the doctor may order an HPV DNA test, which can identify the DNA of 13 high-risk types of HPV in cells taken from the cervix.

There are no HPV screening tests for men.

#### Treatment

Patients with genital warts should *never* use over-the counter-preparations designed to remove common or flat warts from the hands or face.

#### **HPV Infection**

#### WORDS TO KNOW

- **Cervix:** The narrow neck or outlet of a woman's uterus.
- **Cryotherapy:** The use of liquid nitrogen or other forms of extreme cold to treat a skin disorder.
- **Mucous membrane:** Soft tissues that line the nose, throat, stomach, and intestines.

Pap test: A screening test for cervical cancer devised by Giorgios Papanikolaou (1883–1962) in the 1940s.

**Topical:** Referring to a type of medication applied directly to the skin or outside of the body.

Doctors can treat genital warts with various medical or surgical techniques:

- Cryotherapy. Cryotherapy uses liquid nitrogen to freeze the warts. The dead tissue in the wart falls away from the skin beneath in about a week.
- Imiquimod. Imiquimod is a topical cream that gets rid of genital warts by stimulating the body's immune system to fight the virus that causes the warts.
- Podofilox. This is a topical medication available in liquid or gel form that destroys the wart tissue.
- Surgery. The doctor can remove the wart by drying it out with an electric needle and then scraping the tissue with a sharp instrument called a curette. Lasers can also be used to remove genital warts.

Low-grade precancerous changes in the tissue of the female cervix are not usually treated directly, because most of them will eventually go away on their own without developing into cancer. The patient should, however, see the doctor for follow-up Pap smears to make sure that the tissues are returning to normal. High-risk precancerous lesions are removed, usually by surgery, cryotherapy, or laser surgery.

#### Prognosis

The prognosis of sexually transmitted HPV infections depends on the patient's age, number of sexual partners, gender, and the condition of their immune system. Women are significantly more likely than men to develop cancers following HPV infection. However, most people of either sex with normally functioning immune systems who are infected with HPV will clear the infection from their bodies within two years.

#### Prevention

Preventive measures that people can take to lower their risk of HPV infection include:

- Abstaining from sex, or having sex only with an uninfected partner who is faithful.
- Reducing the number of sexual partners.
- Using condoms regularly during sexual intercourse.
- For women, using a new vaccine called Gardasil. Approved by the Food and Drug Administration in 2006, Gardasil is a vaccine that protects against the four types of HPV that cause most cervical cancers and genital warts. The vaccine is recommended for elevenand twelve-year-old girls. It is also recommended for girls and women age thirteen through twenty-six who have not yet been vaccinated or completed the vaccine series. Gardasil works best in girls who have not yet been sexually active. It is given as a series of three shots over a six-month period.

## **The Future**

Researchers are working on developing vaccines that protect against additional types of the HPV virus. Other scientists are studying the possibility that the transmission of HPV could be prevented by applying substances that would kill bacteria or viruses directly to the skin of the genitals or to condoms. Several different gels and creams were in clinical trials as of 2008.

SEE ALSO Warts

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# Huntington Disease

#### Definition

Huntington disease, or HD, is a rare and incurable genetic disorder caused by a defective gene on chromosome 4.

#### Description

HD is a rare but invariably fatal disease caused by an abnormal stretch of DNA in a single gene on chromosome 4. It has been known for centuries; in the Middle Ages it was grouped together with other movement disorders under the name of St. Vitus' dance. Huntington disease got its present name from Dr. George Huntington (1850–1916), an American doctor who published the first medical description of the disease in 1872, one year after completing medical school. Dr. Huntington was the first writer to prove that the disease is inherited; his grandfather and father had both practiced medicine on Long Island and had kept records of a single family with four generations of members with HD.

#### Also Known As

Huntington's chorea, HD, St. Vitus' dance

#### Cause

Defective gene on chromosome 4

#### Symptoms

Jerky, random physical movements followed by mental decline and eventual death

#### Duration

Ten to twenty-five years after symptoms appear

It was not until 1983, however, that geneticists were able to find out which of the twentythree pairs of human chromosomes carries the gene that causes HD. In that year it was discovered that the gene was located somewhere on chromosome 4, although the gene itself was not pinpointed until 1993. In the meantime researchers were studying families living in poor fishing villages along the shores of Lake Maracaibo in Venezuela. This group of families has the highest rate of HD in the world—700 per 100,000 people. Tissue samples from these families helped to locate the gene that causes HD.

## **Demographics**

Huntington disease is uncommon, affecting between four and eight people per 100,000 in the United States and between two and ten per 100,000 in Europe. There are a few countries other than Venezuela with higher than average rates of HD, including Mauritius (forty-six per

100,000 people) and Tasmania (eighteen per 100,000 people). The disease appears to be somewhat more common among people of European ancestry than among Africans or Asians.

Men and women are equally likely to inherit the defective gene, develop the disease, and pass it on to their children. Huntington disease is one of only a few genetic disorders occurring as a dominant trait, that is, to develop the disease a person only requires one abnormal gene (whereas with recessive traits, to cause overt disease, one must have abnormal genes on both pairs of the chromosome). In many cases the first symptoms appear when patients are in their late 30s or early 40s; however, 10 percent of all cases appear in people younger than twenty. This form of the disease is called juvenile HD. The disease has been known to appear in children as young as two years and in adults over eighty years of age. Most patients die between ten and twenty-five years after the first symptoms appear.



*Elderly woman suffering from Huntington's disease.* CONOR CAFFREY / PHOTO RESEARCHERS, INC.

# The House of Love and Faith

The House of Love and Faith, or Casa Hogar Amor y Fe in Spanish, is a clinic in Venezuela run by Dr. Margot de Young for the families near Lake Maracaibo affected by Huntington disease. There are 14,000 descendants of the first known family member who developed the disease in the 1800s living in this area. Many are at risk of developing HD themselves. The fishing villages around the lake have been impoverished by the disease; children often quit school at age seven or eight in order to care for older relatives dying of HD. By the time they are teenagers, many have had children, thus passing on the gene for the disease to the next generation. Because many families in the villages have ten to twelve children, it is not unusual for such families to have five or more children with juvenile HD.

Dr. de Young's clinic, located in a building that was once a bar, has thirty-four beds for patients in the last stages of Huntington disease. The doctor also provides food, medicines, and basic information about health care and nutrition to the patients' family members. The clinic selectively hires its health care workers from among the HD families; they are proud of their work in the small clinic and do their best to keep it a clean and welcoming place.

## **Causes and Symptoms**

Huntington disease is caused by a defective gene on chromosome 4 that produces an abnormal protein. This protein causes the death of nerve cells in various parts of the brain that control movement, cognition (thinking), and behavior. The defect in the gene is a DNA repeat that occurs from thirty-six to 120 times, whereas there are only seven to thirty-five repeats in a normal gene. The larger the number of repeats, the earlier the symptoms of HD are likely to appear; people with more than sixty repeats are likely to develop juvenile HD. Moreover, the number of repeats increases in each successive generation of people with the defective gene; this characteristic of the disease is known as anticipation.

People can inherit the faulty gene from either parent; however, inheriting it from the father appears to speed up the onset of the disease. Most people who develop juvenile HD inherited the defective gene from the father, whereas people who first develop symptoms after age thirty-five are more likely to have inherited the defective gene from their mother.

The symptoms of Huntington disease include physical, mental, and emotional symptoms. In most cases the physical indications of the disease are the first to appear, although some patients have memory problems or emotional disturbances as the earliest symptoms.

- Physical symptoms: uncontrollable fidgeting or sudden jerky movements (chorea); loss of coordination and balance; difficulty changing the direction of the eyes without moving the entire head; uncontrollable facial grimaces; difficulties with speech and swallowing.
- Cognitive symptoms: dementia (loss of memory and ability to make plans or solve problems); disorientation and confusion.

• Emotional changes: depression; personality changes; antisocial behavior; hallucinations; psychosis (complete loss of contact with reality).

Younger patients with juvenile HD may have symptoms resembling those of Parkinson's disease, such as rigid muscles, slow movement, drooling, and frequent falls. Between 30 and 50 percent of these patients also develop seizures.

As the disease progresses, patients gradually lose their ability to walk or stand and may be confined to a wheelchair or completely bedridden. They may become completely stiff or unable to eat; most patients eventually have to be institutionalized.

# Diagnosis

The doctor may order various imaging studies of the brain, such as a computed tomography (CT) scan or magnetic resonance imaging (MRI). Patients with HD will typically show some loss of brain tissue in a specific area of the brain called the caudate nucleus. This part of the brain primarily controls movement but is also involved in learning and memory. It is one of the first parts of the brain to be damaged by the disease.

Genetic testing can be done to confirm the diagnosis of Huntington disease. It involves a blood sample that counts the number of DNA repetitions in the HD gene. People who are at risk for HD can request the test before they develop symptoms. They must undergo several counseling sessions before the test, however, to make sure that they can cope with the results because there is no way to cure HD or slow its appearance.

# Treatment

Treatments for Huntington disease are primarily intended to help patients manage their symptoms, since there is no cure for the disease. Most patients are given several medications, which may include antiseizure drugs, antidepressants, tranquilizers, drugs to control hallucinations and other symptoms of psychosis, and drugs to control involuntary body movements.

Other treatments include physical therapy to help keep the patient's muscles strong and flexible, speech therapy to help with difficulties in

talking clearly, and occupational therapy to help the patient take care of dressing, bathing, and other basic needs as long as possible.

#### Prognosis

There is no cure for HD. Patients die about nineteen years on average after the first symptoms appear, most commonly of pneumonia or another infection, malnutrition from inability to eat, or suicide.

# Prevention

One reason why Huntington disease has not died out among humans is that the earliest symptoms often do not appear until after the affected person has had children and thus conveyed the defective gene into the next generation. People with a parent who has the defective gene should receive genetic counseling before starting a family. Some younger people choose to be tested for the gene before marrying in order to tell whether they will develop HD themselves as well as risk passing on the defective gene to children. People at risk for HD can consider adoption if they wish to start a family, or they can use a form of assisted reproduction in which embryos are screened for the Huntington gene mutation before being implanted in the woman's uterus.

#### The Future

It is not likely that HD will ever be completely wiped out even if everyone who presently might have the defective gene agreed to be tested and further agreed not to have children if they turned out to have the gene. The reason why HD would reappear at some point is that some people can develop the disease even without a family history of HD because of a spontaneous mutation (change in their genetic material).

Researchers are looking in several different directions for a possible cure for Huntington disease. One possibility is gene silencing, a technique that would involve using a short sequence of DNA to block the expression of the DNA repeats on the Huntington gene. Another possibility is a combination of certain drugs used to treat cancer with other drugs used to treat AIDS. This type of drug therapy has not yet been tested in humans, however. Still a third possibility is implanting stem cells in the damaged parts of the brain to replace the nerve cells that have already been destroyed.

## WORDS TO KNOW

- **Anticipation:** A condition in which the symptoms of a genetic disorder appear earlier and earlier in each successive generation.
- **Chorea:** A general term for movement disorders marked by loss of coordination and involuntary motions of the head and limbs.
- **Dementia:** Loss of memory and other mental functions related to thinking or problem-solving.

**Mutation:** A change in the genetic material of an organism.

- **Psychosis:** A severe form of mental illness involving loss of contact with reality.
- **Stem cell:** An unspecialized human cell that has the capacity to form itself into a nerve cell or other type of specialized cell.

#### SEE ALSO Parkinson disease

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# Hutchinson-Gilford Syndrome

# Definition

Hutchinson-Gilford progeria syndrome, or HGPS, is a genetic disorder characterized by premature aging and early death.

# Description

HGPS is a sporadic genetic disorder, which means that it usually occurs at random and occurs in families only rarely. It was first described in 1886 by Jonathan Hutchinson (1828–1913), a British surgeon, and independently reported in 1897 by Hastings Gilford (1861–1941), also a British surgeon.

Children with HGPS look normal at birth but begin to show signs of the disorder during the first two years of life. Their growth slows down; this condition is called failure to thrive or FTT. The child is short and small for his or her age, develops wrinkled skin like that of an elderly person, loses the body fat beneath the skin, and has a fragile-looking body with joints that stick out and easily dislocated hips. The child's facial features are also distinctive, with large eyes, loss of scalp hair, a beaklike nose, thin lips, a small chin, and large ears. Some children have missing teeth or teeth that are late to come in. The child's body ages six to eight times faster than normal aging; however, these children do not

#### Also Known As

Hutchinson-Gilford progeria syndrome, HGPS

#### Cause

Mutations of the LMNA gene

#### Symptoms

Premature aging, hair loss, wrinkled skin, hardening of the arteries, early death

#### Duration

Lifelong; average life expectancy is thirteen years develop certain age-related problems like cataracts or an increased risk of cancer.

The disorder does not interfere with intellectual development or with sitting, standing, and walking. Children with HGPS can go to school with other children and participate in activities with them.

The disease leads to death at an early age from heart attack or stroke caused by premature hardening of the arteries. Children with HGPS frequently experience angina—chest pain caused by an inadequate supply of oxygen to the heart muscle. They also suffer from high blood pressure and enlargement of the heart.

# Demographics

HGPS is a very rare disorder, thought to affect between one in 8 million and one in 4 million children. About 130 cases have been reported in medical journals since the disease was first described in 1886. There were seven children in the United States with progeria in 2005.

As of 2008, 97 percent of reported cases of HGPS had been found in Caucasians. The reason

for this racial disparity is not yet known; since some researchers think that the disorder is misdiagnosed in some cases, it is possible that the racial difference is at least partly a matter of reporting.

HGPS affects boys slightly more often than girls; the gender ratio is about 1.5 boys for every girl.

For many years it was thought that HGPS does not run in families. Since the early 2000s, however, two families have been identified with more than one child affected by the disorder. The first is a family in India with five children with HGPS, first described by a pediatrician in Calcutta in 2005. The other is a family in Belgium with two children with HGPS that was diagnosed in 2006.

# **Causes and Symptoms**

Hutchinson-Gilford progeria syndrome is caused by a mutation in a gene on chromosome 1 called the LMNA gene. This gene tells cells how to



John Tacket, age fifteen, has Hutchinson-Gilford syndrome, or progeria, a rare disease of accelerated aging. AP IMAGES.

# Coping with Unfair Suffering

Although Hutchinson-Gilford syndrome is an extremely rare disorder, it played a role in the writing of a well-known book on the meaning of human suffering. When Bad Things Happen to Good People was first published in 1981 and reissued in 2001 with a new preface. The author, Harold S. Kushner, was moved to write the book following the death of his son Aaron in 1977 at the age of fourteen. Rabbi Kushner had been the spiritual leader of Temple Israel in Natick, Massachusetts, when his son was diagnosed with progeria at the age of three in 1966. He promised his dying son that he would tell his story so that Aaron would not be forgotten. Four years after Aaron's death, the book was finally ready for publication.

Written in a warm, accessible tone, *When Bad Things Happen to Good People* quickly became a best seller, bringing comfort to literally millions of people struggling with sorrow and loss. The book was eventually translated into fourteen languages and still prompts readers to send thank-you notes to its author. Rabbi Kushner is not, however, concerned with being remembered as an author. He told an interviewer in 2003, "I want to be remembered that, when my son was dying and in pain, I could make him laugh." make a protein called lamin A, which helps to shape the cell nucleus inside the cell. The defective gene involved in HGPS cannot give the cell proper instructions for making lamin A. As a result, the cell nucleus develops into a strange and twisted shape rather than the normal round shape. It is not known, however, just how the unstable shape of the cell nucleus is related to the characteristic symptoms of HGPS.

# Diagnosis

Children with HGPS are usually diagnosed around two years of age, when the changes in their skin, their distinctive facial features, and their failure to grow normally become apparent. The diagnosis can be confirmed by a blood test that was developed in 2003 after the gene that causes the disorder was first identified.

The doctor may also take a small sample of skin to examine it under the microscope for the changes that indicate HGPS, but this test is not necessary to diagnose the disorder.

# Treatment

There is no treatment that can cure HGPS. Therapy is intended to give the child as normal a life as possible. Some children are given a daily aspirin to counteract the risk of heart attack or stroke, and doctors commonly recommend a high-calorie diet to help them gain weight. Chil-

dren with HGPS may also benefit from physical therapy to keep their muscles and joints from weakening.

Children with HGPS must see the doctor periodically to have the condition of their heart and major blood vessels checked and to have their food intake adjusted when necessary.

# Prognosis

HGPS is invariably fatal; 90 percent of children with the disorder die of heart attacks or stroke. The average life expectancy is thirteen years,

## WORDS TO KNOW

**Angina:** Chest pain caused by an inadequate supply of blood to the heart muscle.

Failure to thrive: A term used to describe children whose present weight or rate of weight gain is markedly lower than that of other children of their age and sex. **Progeria:** A disease characterized by abnormally rapid aging. The term can be used to refer specifically to Hutchinson-Gilford syndrome or to a group of diseases characterized by accelerated aging.

Werner syndrome: Another genetic disease characterized by accelerated aging.

although some children die as young as six or seven. The longest-lived person with HGPS died at twenty-nine.

#### Prevention

Since HGPS is a genetic disorder, there is no known way to prevent it.

## The Future

HGPS is of interest to researchers, along with such other disorders of accelerated aging as Werner syndrome, because they think these diseases may hold clues to the normal process of human aging. In regard to a possible cure for HGPS, a clinical trial of a drug called lonafarnib, originally developed to treat cancer, began in May 2007. Some researchers are experimenting with growth hormone as a possible treatment for HGPS, but the results have not been encouraging.

#### SEE ALSO Heart attack; Stroke

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# Hydrocephalus

# Definition

Hydrocephalus is a condition in which the flow of cerebrospinal fluid (CSF) in the central nervous system is interrupted or blocked. CSF is the liquid that circulates between the layers of tissue that cover the brain, within the ventricles (hollow cavities) of the brain, and around the spinal cord. It serves to cushion the structures of the central nervous system, deliver nutrients to the brain, and regulate the amount of blood within the brain. In normal circumstances, the CSF moves within the ventricles in the brain, exits through closed spaces at the base of the brain, flows over the surface of the brain and spinal cord, and is then reabsorbed into the bloodstream.

Hydrocephalus develops when there is an imbalance between the production of CSF and its reabsorption or when its flow is blocked. The cerebrospinal fluid builds up inside the brain, putting pressure on the tissues of the central nervous system and causing symptoms ranging from visual disturbances and headache to mental disturbances and difficulty walking.

Also Known As Water on the brain

#### Cause

Birth defects, head injuries, brain tumors, stroke, infections, bleeding in the brain

#### Symptoms

Swollen head (infants); headache, nausea and vomiting, loss of balance, problems with memory

#### Duration

Months to years unless corrected

Doctors classify hydrocephalus into several different categories:

- Congenital. This type of hydrocephalus is present at birth and may be caused by genetic disorders or problems that occur during the baby's development before birth.
- Acquired. Acquired hydrocephalus develops later in life as the result of brain tumors, head injuries, infections of the brain, or other brain disorders.
- Communicating. This type of hydrocephalus is one in which the CSF can flow between the ventricles of the brain but is blocked from leaving the brain.
- Noncommunicating or obstructive. In this type of hydrocephalus, the CSF cannot flow freely among the ventricles inside the brain.
- Normal pressure hydrocephalus (NPH). This is a form of communicating hydrocephalus most commonly found in the elderly. It is a condition in which CSF builds up within the ventricles of the brain.



Mother carrying her six-monthold son, who has bydrocephalus, in Calcutta, India. AP IMAGES.

# Description

The symptoms and course of hydrocephalus vary widely depending on the person's age, the cause of the condition, and its severity. In infants, the bony plates that form the skull have not yet completed their joining together. This incomplete development of the skull allows the infant's head to expand from the pressure of the CSF, sometimes very rapidly. The baby may vomit, sleep a lot, be irritable, or have seizures. In older children and adults, the skull has already completed its development and the buildup of CSF results in increased pressure on the tissues of the brain and spinal cord. Hydrocephalus in these age groups is more likely to produce such symptoms as headaches, double vision, vomiting, problems with balance or coordination, drowsiness, personality changes, or other signs of damage to the central nervous system.

# An Unusual Case of Hydrocephalus

In July 2007, the British medical journal *The Lancet* published a report by three French surgeons who had treated a forty-four-year-old man in a Marseille hospital for weakness in his left leg. When the doctors performed some imaging studies of the man's brain, they were amazed to find that the ventricles in his brain had filled with cerebrospinal fluid (CSF) to the point that his brain had been crushed against the sides of his skull. There was very little brain tissue left. The chief surgeon was quoted as saying, "The images [from the scans] were most unusual...the brain was virtually absent."

The surgeons thought that the man's condition was the result of an operation he had had at the age of six months to treat hydrocephalus. Although the man had an IQ of 75, somewhat below normal, he had led a normal life without any unusual medical symptoms until his leg disorder. He was married and the father of two children and was employed as a civil servant. The surgeons treated the man by inserting a new shunt to drain the excess CSF, which relieved his leg symptoms and allowed him to return to work. Elderly adults with normal-pressure hydrocephalus often have difficulties with bladder control and movement as well as dementia. Because these symptoms are also found in such disorders as Parkinson disease or Alzheimer disease, many older adults with NPH are never properly diagnosed or treated.

# Demographics

The exact number of people with hydrocephalus in the United States is not known because the disorder has so many possible causes-particularly acquired hydrocephalus-and because the diagnosis is often missed in elderly patients. The National Institutes of Health (NIH) estimates that there are 700,000 children and adults living with hydrocephalus in the United States. It is the leading cause of brain surgery for American children and costs the nation about \$1 billion every year in health care expenses. The disorder is most common in the very young and the very old. About three babies in every 1,000 are diagnosed with congenital hydrocephalus. About 60 percent of cases of acquired hydrocephalus occur in children, with the remaining 40 percent in adults.

As far as is known, hydrocephalus is equally common in both sexes and in all races and ethnic groups.

Risk factors for hydrocephalus in infants and young children include:

- Premature birth. Prematurity increases the risk of bleeding into the brain.
- Spina bifida. This is a condition in which the spinal column fails to close completely over the spinal cord.
- An infection within the mother's uterus.

# **Causes and Symptoms**

The causes of hydrocephalus range from genetic disorders and incomplete development before birth to brain tumors, head injuries, infectious diseases that affect the brain, and bleeding in the brain. The symptoms of hydrocephalus depend partly on the patient's age:

- Infants: Enlargement of the skull; bulging of the soft spot at the top of the skull; veins in the scalp are enlarged; baby feeds poorly, vomits, has seizures, sleeps a great deal, or has eyes that look downward much of the time ("sunsetting").
- Children: Headache; nausea; vomiting; fever; blurred or double vision; unstable balance; irritability; sleepiness; delayed progress in walking or talking; poor coordination; change in personality; difficulty staying awake or waking from sleep.
- Adults: Headache; constant drowsiness; loss of ability to think clearly or concentrate; difficulty walking; personality changes and loss of social skills. Job performance is often affected.
- Elderly adults: Loss of coordination or balance; shuffling gait, memory loss; headache; or bladder control problems.

#### Diagnosis

The specific diagnostic techniques that the doctor will use depend on the person's age and recent medical history. The doctor will note the specific symptoms and when they first appeared. If the patient is an infant, his or her head will be measured and compared to the normal range for babies of the same sex and age. A head larger than 97 percent of the heads of normal children usually indicates hydrocephalus.

Older children and adult patients will usually be referred to a neurologist (a doctor who specializes in treating disorders of the central nervous system) for a complete evaluation of his or her vision, memory, coordination, and other functions that may be affected by hydrocephalus.

The neurologist will order one or more imaging studies of the brain in order to determine whether the hydrocephalus is communicating or noncommunicating and whether other abnormalities of the brain are present. Ultrasound is often used to evaluate hydrocephalus in infants, and computed tomography (CT) scans or magnetic resonance imaging (MRI) is used for older children and adults.

Normal-pressure hydrocephalus is diagnosed by lumbar puncture (spinal tap) followed by withdrawal of some of the cerebrospinal fluid. If the patient has NPH, their symptoms will usually improve after the fluid is removed. This test is known as the Fisher test.

#### Treatment

The usual treatment of hydrocephalus, whatever its cause, is the surgical insertion of a shunt system. A shunt is a flexible plastic tube that carries extra CSF away from the brain. The shunt system consists of the shunt itself, a valve that keeps the CSF flowing in the correct direction, and a long thin tube called a catheter. The shunt is inserted into one of the brain's ventricles. The catheter and valve are attached to it, and the catheter tubing is threaded underneath the skin to another part of the body (usually the heart or the abdomen) where the excess CSF can be absorbed. The shunt system needs periodic replacement in children as they grow or in adults if the tubing becomes blocked or infected.

A few people with noncommunicating hydrocephalus can be treated by surgery on the third of the brain's four ventricles. In this procedure, the surgeon uses a miniature camera and instrument to locate the third ventricle and cut a small hole in its floor. This hole allows the CSF to bypass the blockage between the ventricles and flow toward its normal outlet from the brain.

#### Prognosis

Hydrocephalus cannot be cured. The outcome for a given patient is difficult to predict, as the condition has so many different possible causes. The insertion of a shunt system carries some risk of further brain damage. An estimated 50 percent of all shunts fail within two years, requiring further surgery to replace them. Since 1980, however, death rates associated with hydrocephalus have decreased from 54 percent to 5 percent; and intellectual disability in children with hydrocephalus has decreased from 62 percent to 30 percent.

#### Prevention

The best way to prevent hydrocephalus in newborns is to take steps to reduce the risk of premature birth and to protect infants and small children against head injuries. In addition, vaccinating children against meningitis—a type of infection that can cause hydrocephalus—offers further protection.

#### The Future

Some possible new treatments for hydrocephalus as well as various improvements in shunt systems are currently being studied in clinical

## WORDS TO KNOW

**Dementia:** Loss of memory and other mental functions related to thinking or problem-solving.

**Sunsetting:** A term used to describe a downward focusing of the eyes.

**Shunt:** A flexible plastic tube inserted by a surgeon to drain cerebrospinal fluid from the brain and redirect it to another part of the body.

**Ventricle:** One of four hollow spaces or cavities in the brain that hold cerebrospinal fluid.

trials. As of 2008 the NIH was sponsoring thirty-two separate trials for these treatments.

**SEE ALSO** Alzheimer disease; Brain tumors; Meningitis; Prematurity; Spina bifida; Stroke

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# Hypercholesterolemia

# Definition

Hypercholesterolemia is the medical term for high blood cholesterol levels. It is not a disease as such but a condition that raises a person's risk of coronary heart disease, stroke, and other disorders of the circulatory system.

# Description

Cholesterol is a waxy or fatty substance that the human body produces normally. About 75 percent of the cholesterol in the body is made by the liver and other cells; the remaining 25 percent comes from food. A certain amount of cholesterol is necessary to maintain the function of cell membranes; thus cholesterol is present in the walls of all body cells, including those in the skin, muscle tissue, nervous system, digestive tract, and other parts of the body.

The body also needs cholesterol to make bile (a substance produced in the liver that helps to digest fat), hormones, and vitamin D. This cholesterol is carried in the bloodstream attached to protein molecules. These combinations of cholesterol and protein molecules are called lipoproteins. If more cholesterol is made than is needed for the body's functions, the waxy cholesterol may form deposits on the inner walls of arteries known as plaques.

Fatty plaque deposits are particularly likely to build up in the arteries that supply the heart with blood. These blood vessels are known as the

Also Known As High blood cholesterol

#### Cause

Genetic factors, obesity, high-fat diet

#### Symptoms

None; requires a blood test to detect

#### Duration

Years

coronary arteries. Plaque deposits can become thick enough to partially block the coronary arteries. If the deposits remain in place, they eventually cause the arteries to stiffen or harden—a condition known as atherosclerosis. If the arteries become too narrow because of the plaques, they cannot carry enough blood to the heart to meet the needs of the heart muscle for oxygen. The oxygen-starved muscle may then produce a kind of chest pain known as angina. The fatty plaques can also come loose from the walls of the artery, resulting in the formation of a clot, a complete blockage of the coronary artery, and a heart attack.

It is important to understand that there are three different types of cholesterol and lipoproteins in the human body:



- Low-density lipoprotein (LDL). Often called "bad" cholesterol, LDL is the type of cholesterol that forms plaques on the walls of the coronary arteries.
- High-density lipoprotein (HDL). The "good" cholesterol, HDL picks up LDL and takes it back to the liver. Between 25 and 32 percent of the body's cholesterol is HDL.
- Very low-density lipoprotein (VLDL). This type of cholesterol contains the highest levels of triglycerides (a type of fat) attached to its protein molecules. VLDL is converted in the bloodstream to LDL and can increase the size of LDL particles, thus speeding up the formation of plaques and atherosclerosis.

# **Demographics**

High blood cholesterol levels are largely an adult health problem. Women in the United States before menopause usually have lower blood cholesterol levels than men of the same age. As women and men age, however, their blood cholesterol levels rise until about sixty to sixty-five years of age. After about age fifty, women often have higher total cholesterol levels than men of the same age.

Race and ethnicity appear to affect the rates of hypercholesterolemia in the United States. According to a government health survey carried out Cross section of an artery around the heart. The inside is coated with plaque, making the opening smaller. © PHOTOTAKE INC. / ALAMY. in the 1990s, Caucasian adults are more likely (19 percent) to have high blood cholesterol levels than Hispanics (15 percent) or African Americans (16 percent).

Other risk factors for high blood cholesterol include:

- Smoking. Smoking damages the walls of the coronary arteries, making it easier for plaques to form. It also lowers the level of HDL cholesterol.
- Obesity.
- A high-fat diet. Such high-fat foods as red meat, eggs, and full-fat milk and cheese raise blood cholesterol levels.
- Lack of exercise. Exercise helps to raise HDL levels and lower LDL levels.
- High blood pressure. Like smoking, high blood pressure damages the walls of the coronary arteries.
- Diabetes. High levels of blood sugar raise LDL levels and lower HDL levels.
- Family history of heart disease. A parent or sibling who developed heart disease before age fifty-five places a person with high cholesterol levels at a greater than average risk of developing heart disease.
- Emotional stress. Several studies have shown that high stress levels for long periods of time raise blood cholesterol levels.

# **Causes and Symptoms**

The basic cause of high blood cholesterol levels is a combination of genetic factors and lifestyle factors, particularly diet. There is one specific form of hypercholesterolemia called familial hypercholesterolemia that affects about one person in every 500 in the United States. Familial hypercholesterolemia is caused by a mutation in one specific gene known as the LDLR gene.

Genetic factors, however, also affect other people's risk of hypercholesterolemia. As of 2008 no other specific genes had been associated with high blood cholesterol levels in the general population; researchers think that there are probably several such genes rather than only one. These genetic factors contribute to high cholesterol levels either by interfering with the body's ability to remove LDL cholesterol from the bloodstream or by allowing the liver to produce too much cholesterol. A person can have high blood cholesterol levels without any noticeable symptoms. Because of this fact, the National Cholesterol Education Program (NCEP) guidelines suggest that everyone aged twenty years and older should have their blood cholesterol level measured at least once every five years.

## Diagnosis

Blood cholesterol levels are measured by a blood test taken early in the morning after nine to twelve hours of fasting. The doctor will ask the patient about a family history of high cholesterol or heart disease as well as drawing the blood, since high cholesterol levels can be hereditary.

The blood cholesterol test measures total blood cholesterol, LDL, HDL, and triglyceride levels using units called milligrams per deciliter (mg/dL).

- Total cholesterol: Less than 200 mg/dL is a desirable level that lowers a person's risk for heart disease. A cholesterol level of 200 mg/dL or greater increases the risk. A level of 240 mg/dL and above is considered high blood cholesterol. The risk of heart disease at this level is twice that of a person whose total cholesterol level is 200 mg/dL.
- LDL: Less than 100 mg/dL is considered the best level; a level of 100–129 mg/dL is good; a level of 130–159 mg/dL is borderline high; a level of 160–189 mg/dL is high; and a level of 190 mg/dL and above is very high.
- HDL: A level below 40 mg/dL is considered a major risk factor for heart disease; a level between 40 and 59 mg/dL is better; and a level above 60 mg/dL is considered protective against heart disease.
- Triglycerides: Less than 150 mg/dL is normal; a level of 150–199 mg/dL is borderline high; a level of 200–499 mg/dL is high; and a level of 500 mg/dL or above is very high.

#### Treatment

Treatment for hypercholesterolemia begins with lifestyle changes, including following strict dietary guidelines and increasing one's amount of daily exercise. There is some evidence that a vegetarian diet is beneficial.

In addition to lifestyle changes, the patient's doctor may recommend one or more medications to lower LDL and/or triglyceride levels. The most common types of drugs used to control hypercholesterolemia are:

- Statins. These are drugs that block the liver from using a substance it needs to make cholesterol. As the level of cholesterol in the liver drops, the liver begins to remove excess cholesterol from the bloodstream.
- Bile acid-binding resins. These are drugs that work by prompting the liver to make more bile acid; to do this, the liver needs to draw cholesterol from the blood.
- Cholesterol absorption inhibitors. These medications work by limiting the amount of cholesterol that the small intestine can absorb from food.
- Fibrates. These are drugs that speed up the removal of triglycerides from the bloodstream.
- Niaspan. Niaspan is a prescription form of niacin (a B vitamin) that works by limiting the liver's ability to produce VLDL and LDL cholesterol.

# Prognosis

The prognosis of hypercholesterolemia depends on the person's age, sex, family history, and willingness to follow a treatment program. The statins in particular have greatly improved a person's ability to lower his or her risk of coronary heart disease. The United States Preventive Services Task Force (USPSTF) has estimated that five to seven years of treatment with statins can lower the risk of heart disease by 30 percent.

# Prevention

People cannot change their age, sex, genetic factors, or family history that may increase their risk of high cholesterol levels, but they can manage their risk by getting plenty of exercise, keeping their weight at a healthy level, quitting smoking, and eating foods that help to lower LDL levels.

Specific dietary recommendations include:

- Eating foods that are low in saturated fats.
- Keeping cholesterol intake from foods below 200 milligrams per day. One egg, for example, contains about 210 milligrams of cholesterol. Eating lean meats, drinking skim instead of whole milk, and using egg substitutes are good ways to lower one's intake of cholesterol from foods.

# WORDS TO KNOW

- **Angina:** Chest pain caused by inadequate supply of oxygen to the heart muscles.
- Atherosclerosis: Stiffening or hardening of the arteries caused by the formation of plaques within the arteries.
- **Cholesterol:** A fatty substance produced naturally by the body that is found in the membranes of all body cells and is carried by the blood.
- **Plaque:** A deposit of cholesterol along the inside wall of an artery.

Triglyceride: A type of fat made in the body.

- Eating whole-grain breads.
- Adding more servings of fruits and vegetables to the diet. These foods are rich in fiber, which can lower blood cholesterol levels.
- Eating more fish. Some types of fish, such as cod, halibut, and tuna, are lower in fat and cholesterol than poultry or red meat.
- Keeping one's alcohol intake moderate.

# **The Future**

Researchers are presently focusing on the genetic factors involved in hypercholesterolemia as well as potential new treatments for it. As of 2008, the National Institutes of Health (NIH) was conducting almost 400 separate studies on high blood cholesterol, ranging from clinical trials of new statins to studies of statins in combination with other drugs, and several experimental drugs.

SEE ALSO Coronary artery disease; Heart attack; Stroke

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# Hyperopia

# Definition

Hyperopia is one of several eye conditions called refractive errors, which means that light entering the eye is not properly focused on the retina (the light-sensitive layer of tissue at the back of the eyeball). It is not a disease of the eye in the strict sense.

# Description

Hyperopia, or farsightedness, is a condition that develops when a person's eyeball is abnormally short from front to back, or when the cornea (the clear front portion of the eyeball) is abnormally flat. In a normal eye, light entering the eye through the cornea is focused by the lens of the eye on the retina. In hyperopia, the abnormal shortness of the eye or the flatness of the cornea causes the lens to focus images behind the retina. This incorrect focus means that objects at a distance can be seen more clearly than those that are close to the viewer. If the hyperopia is severe, the person may be able to see clearly only objects that are quite far away.

# Demographics

Hyperopia is a common refractive error in the general population, affecting about 25 percent of the general population. In addition, the condition tends to run in families. Hyperopia is often combined with astigmatism, another type of refractive error caused by irregularities in the curvature of the cornea or the lens of the eye.

Most babies are mildly hyperopic at birth. Hyperopia in children is usually less severe than hyperopia in adults, partly because the eyeball in many children lengthens as they grow older and allows the eye to focus

#### Also Known As Farsightedness

#### Cause

Short eyeball or overly flat cornea

#### Symptoms

Blurry near vision; sometimes blurry far vision as well

#### Duration

May be lifelong





normally. It is thought that between 6 and 9 percent of children in the United States may have mild hyperopia. Boys and girls are equally affected. There are, however, racial and ethnic differences, with Native Americans, African Americans, and Pacific Islanders having higher than average rates of hyperopia.

# **Preventive Eye Care**

Protection of one's vision is an important part of preventive health care. Eye doctors recommend the following schedule of eye examinations for children and adults:

- Children: should have their eyes tested by their pediatrician or an ophthalmologist between birth and three months; between six months and one year; at three years; and at five years.
- Older children and adolescents: should be seen yearly if they wear corrective lenses, have other eye problems, or have diseases like diabetes that affect the eyes.
- Adults between twenty and thirty-nine: should have the eyes checked every three to five years.
- Adults over thirty-nine who do not wear glasses and are at low risk of eye disease: should be checked every two years between forty and sixty-four, and every year after age sixty-five.
- Adults over thirty-nine who do wear eyeglasses or contact lenses: should have an eye checkup every year.

People who notice blurriness, pain, or any other visual problem should make an appointment with their eye doctor as soon as possible even if they recently had an eye checkup.

There is a condition similar to hyperopia called presbyopia that appears in middle-aged adults. Presbyopia is a type of farsightedness that develops because the lens of the eye becomes less flexible with age and cannot change its shape as easily when the person is trying to focus on near objects (usually reading materials). Most people over forty will develop some degree of presbyopia. Hyperopia that went unnoticed during a person's younger years may become apparent in middle age, when the person begins to develop presbyopia as well.

# **Causes and Symptoms**

In addition to a short eyeball or flatter cornea, hyperopia can be caused in some people by abnormal development of the eye or by trauma to the eye. In a very few cases, hyperopia may be related to disorders of the nervous system or to medications that affect the eye's ability to focus. In general, genetic factors are thought to play a more important role in hyperopia than environmental factors or personal history.

Hyperopia in younger children may not cause noticeable symptoms. Older children and adults, however, will often develop the following symptoms:

- Having to squint while reading.
- Frequent blinking and difficulty focusing on close objects.
- Red or teary eyes, or burning or aching in the eyes.
- Blurry vision.
- Headaches or general discomfort in the eye after a long period of reading, writing, or doing other close work.

People who have these symptoms should make an appointment with an optometrist (an eye care professional who is trained to diagnose refractive errors) or an ophthalmologist (a doctor who specializes in diagnosing diseases of the eye) to find out whether they need corrective lenses.

#### Diagnosis

Hyperopia and other refractive errors are evaluated by a series of vision tests. After the examiner takes a history of the patient's symptoms (including a family history of eye problems), the patient is usually asked to read the letters on an eye chart. The examiner may also shine lights into the eyes or administer eye drops that allow him or her to see all the structures inside the eye clearly.

To measure the strength of the lens needed to correct the patient's hyperopia, the examiner uses a device called a photopter (or refractor). The photopter is placed in front of the patient's eyes, and the examiner moves various lenses in and out of the device while the patient reads letters on an eye chart located 20 feet (6 meters) away.

#### Treatment

Screening for and treatment of hyperopia in school-age children is important because significant hyperopia can lead to strabismus (inability of the eyes to work together) or amblyopia, a condition in which there is poor vision in one eye that is not caused by disease. In addition, uncorrected hyperopia can lead to problems in school, including learning disorders and loss of interest in reading.

Hyperopia can be treated nonsurgically by prescription eyeglasses or contact lenses, which are prescribed by the optometrist or ophthalmologist but made and fitted by an optician. There are also surgical options for people who dislike glasses or contact lenses. The two most common surgical procedures for hyperopia involve reshaping the cornea with a laser or implanting an artificial lens in the front of the eye. Reshaping the cornea works better if the refractive error is only low to moderate. Patients with a high degree of refractive error generally do better with lens implantation.

There are drawbacks to surgical correction for refractive errors, however. These include the risks of infection, development of haze in the cornea, or dry eyes. In some cases the surgeon may need to perform a second operation if the first one either overcorrected or undercorrected the shape of the patient's cornea.

It is important for a patient diagnosed with hyperopia to discuss all the treatment options with the optometrist or ophthalmologist, as no

# WORDS TO KNOW

- **Amblyopia:** Dimness of sight in one eye without any change in the structure of the eye.
- **Astigmatism:** A refractive error caused by irregularities in the shape of the cornea or the lens of the eye.
- **Cornea:** The transparent front part of the eye where light enters the eye.
- **Ophthalmologist:** A doctor who specializes in diagnosing and treating eye disorders and can perform eye surgery.
- **Optician:** An eye care professional who fills prescriptions for eyeglasses and corrective lenses.

- **Optometrist:** An eye care professional who diagnoses refractive errors and other eye problems and prescribes corrective lenses.
- **Photopter:** A device positioned in front of a patient's eyes during an eye examination that allows the examiner to place various lenses in front of the eyes to determine the strength of corrective lenses required.
- **Presbyopia:** Age-related farsightedness caused by loss of flexibility in the lens of the eye.
- **Refractive error:** A general term for vision problems caused by the eye's inability to focus light correctly.
- **Strabismus:** A condition in which the eyes are not properly aligned with each other.

two people will have exactly the same degree of farsightedness or the same lifestyle.

#### Prognosis

Most patients with hyperopia do well after being fitted with corrective lenses or having eye surgery. Hyperopia caused by the shape of the eyeball or the cornea does not get worse with age and is unlikely to lead to vision loss.

#### Prevention

Hyperopia is largely a matter of heredity and cannot be prevented. People can, however, prevent strabismus or other complications of hyperopia by visual screening in childhood and regular eye checkups at all ages.

#### The Future

It is possible that laser treatment and other types of vision surgery will be further refined in the future and have fewer risks or side effects.

SEE ALSO Astigmatism; Myopia; Strabismus

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# **m** Hypertension

# Definition

Hypertension, or high blood pressure (HBP), is a condition in which a person's blood pressure is higher than is healthy over a period of time. Blood pressure normally rises and falls over the course of a day; it also

rises when a person is anxious or is exercising. Doctors usually take blood pressure readings during two different office visits before diagnosing hypertension.

Blood pressure is measured in millimeters of mercury (mm Hg) because the sphygmomanometer—the device most commonly used to measure blood pressure—contains a column of mercury that rises and falls as the doctor inflates and deflates a cuff around the patient's arm. The first of two numbers in a blood pressure measurement is the systolic blood pressure. It is the peak blood pressure in the patient's arteries. The second measurement is the diastolic blood pressure and represents the lowest blood pressure, which occurs when the heart is resting between beats.

Doctors use the following values, measured in millimeters of mercury (mm Hg), when evaluating a patient for hypertension:

- Normal blood pressure: systolic below 120 mm Hg, diastolic below 80.
- Pre-hypertension: systolic between 120 and 139 mm Hg, diastolic between 80 and 99.
- Stage 1 hypertension: systolic between 140 and 159 mm Hg, diastolic between 90 and 99.
- Stage 2 hypertension: systolic 160 mm Hg or higher, diastolic 100 or higher.

Doctors also distinguish between primary (or essential) hypertension and secondary hypertension. Essential hypertension, which accounts for about 95 percent of cases in American adults, is high blood pressure that develops without any apparent cause. Secondary hypertension, which accounts for the remaining 5 percent, is caused by other diseases or conditions, including pregnancy, abnormalities in the shape of the aorta, kidney disease, alcoholism, thyroid disease, the use of birth control pills, and tumors of the adrenal gland.

#### Description

Hypertension is considered a silent disease because a person can have it for years without any noticeable symptoms. Although some people develop nosebleeds, headaches, dizziness, nausea, or changes in their vision as a result of hypertension, the majority of those diagnosed with high blood pressure do not know they have a problem until their doctor checks their blood pressure during a routine physical exam. Sadly, some

#### Also Known As

High blood pressure, HBP

#### Cause

Unknown (essential hypertension); medications, eating habits, pregnancy, diseases (secondary)

#### Symptoms

Often none at all; chest pain, tiredness, ringing in the ears, vision changes

#### Duration

Years

#### Hypertension



Image of the retina, the lining of the back of eye. In hypertension, the small arteries that are visible get thicker, and push on the veins so they appear nipped as they cross the arteries. © MEDICAL-ON-LINE / ALAMY.

people do not know they have high blood pressure until they have a stroke or heart attack.

Untreated hypertension can damage the heart and other organs:

- High blood pressure can lead to heart failure by causing the heart to enlarge or grow weaker.
- Hypertension can lead to stroke by causing a bulge or weak spot along the wall of an artery known as an aneurysm.
- Blood vessels in the eye can burst or bleed as a result of high blood pressure, leading to loss of vision.
- High blood pressure can cause the blood vessels in the kidneys to become too narrow, leading eventually to kidney failure. Narrowing of the blood vessels in the heart may eventually cause a heart attack.

#### **Demographics**

Hypertension is almost entirely a disorder of adults; it affects only 1–3 percent of children and teenagers. About 35 percent of adults in the United States have high blood pressure. Worldwide, hypertension is a leading cause of illness and death; it is the most important factor that people can control in regard to their risk of heart failure, heart disease, stroke, and kidney failure.

Risk factors for hypertension include:

- Age: As people get older, their arteries tend to stiffen, thus contributing to high blood pressure.
- Race: African Americans develop hypertension at higher rates than members of other races in the United States. They also develop high blood pressure at younger ages.
- Sex: Men are more likely to develop hypertension than women.
- Obesity: People with a body mass index (BMI) over thirty are at increased risk of high blood pressure.
- Family history: Hypertension is known to run in families.
- Lack of exercise.
- Drugs of abuse: Cocaine, amphetamines, and heavy alcohol use all increase a person's risk of hypertension.

# **Causes and Symptoms**

The causes of essential hypertension are not known.

Causes of secondary hypertension include:

- Chronic kidney disease
- Tumors or other diseases affecting the adrenal gland
- An unusually narrow aorta (Some people are born with this condition.)
- Pregnancy
- Use of birth control pills
- Thyroid disorders

# Diagnosis

Hypertension is diagnosed in the doctor's office by the use of a sphygmomanometer. The doctor places a cuff around the patient's upper arm and inflates it while listening through a stethoscope. The cuff is inflated until the air pressure squeezes the large artery in the upper arm shut. The doctor then releases pressure in the cuff until he or she can start to hear the sounds of the patient's pulse. This is the systolic pressure. The doctor then continues to release pressure until the sounds disappear. This second reading is the diastolic pressure. In addition to measuring the patient's blood pressure, the doctor may order additional tests to look for possible causes of the hypertension or signs of damage to other organs:

- Blood and urine tests. These can be done to look for evidence of kidney or thyroid disease or to measure the levels of cholesterol in the patient's blood.
- X-ray studies of the chest or the kidneys.
- Electrocardiogram (ECG or EKG). This test measures the electrical activity of the heart and may be done to evaluate the condition of the patient's heart muscle.

#### Treatment

There is no cure for hypertension. Treatment for the disorder comprises a combination of lifestyle changes and medications. Some people can lower their blood pressure by losing weight and getting more exercise, but most need to take medications to keep their blood pressure within the normal range.

The doctor may prescribe one or a combination of medications to control the patient's blood pressure. In most cases the patient will be asked to see the doctor every three to four months to see whether the drugs or their dosage levels need to be adjusted. Although these drugs can produce side effects, the patient should not stop taking them without consulting the doctor. The various types of drugs given to control hypertension include:

- Diuretics. Sometimes called water pills, diuretics increase salt and urine output. They are often prescribed in combination with other types of pills.
- ACE inhibitors. These drugs work to lower blood pressure by blocking the production of a chemical that causes blood vessels to tighten.
- Calcium channel blockers. These drugs work by reducing the force of the contraction of the heart muscle.
- Beta blockers. Drugs in this group slow down the heart rate. They are often prescribed for people who have had a heart attack or a history of heart disease.
- Vasodilators. These drugs cause blood vessels to open up, which lowers blood pressure. They may be given intravenously if the patient's blood pressure has risen very high very quickly.
• Alpha blockers. Drugs in this group block nerve impulses that cause blood vessels to tighten up. The blood can then move more freely through the blood vessels, thus lowering blood pressure.

# Prognosis

Most patients with hypertension can keep their blood pressure at a healthy level provided they follow their doctor's recommendations about diet and exercise and take their blood pressure medications. They should watch their blood pressure carefully as they age because hypertension tends to get worse as people get older. Even mild hypertension, if untreated, increases a person's risk of heart disease by 30 percent and kidney damage by 50 percent within eight to ten years after it starts.

# Prevention

Hypertension is a lifelong disorder that requires long-term commitment to healthy lifestyle changes and regular use of prescribed medications.

Specific lifestyle changes recommended by the National Institutes of Health (NIH) for controlling hypertension include:

- Eating a healthful diet. A good place to start is the Dietary Approaches to Stop Hypertension (DASH) diet, which emphasizes eating more fruits, vegetables, and low-fat dairy products, and cutting back on salt and alcohol.
- Get regular physical exercise, preferably at least thirty minutes a day.
- Stop smoking (or do not start).
- Learn to manage emotional stress effectively; relaxation techniques, biofeedback, meditation, or yoga work well for many people.
- Lose weight if the doctor recommends it.
- Avoid cocaine, heavy drinking, and other recreational drugs that raise blood pressure.
- Have blood pressure checked regularly and take all medications for hypertension on a daily basis even if you feel fine.

# The Future

The rates of hypertension in all developed countries are expected to rise over the next several decades as populations get older and the rates of obesity continue to increase. A major difficulty in treating hypertension is that

# WORDS TO KNOW

- **Aneurysm:** A weak or thin spot on the wall of an artery.
- **Aorta:** The large artery that carries blood away from the heart to be distributed to the rest of the body.
- **Diastolic blood pressure:** The blood pressure when the heart is resting between beats.
- **Essential hypertension:** High blood pressure that is not caused by medications, pregnancy, or another disease.
- **Sphygmomanometer:** The device used to measure blood pressure. It consists of an inflatable cuff that compresses an artery in the arm. The doctor listens through a stethoscope as the air pressure in the cuff is released in order to measure the blood pressure.
- **Systolic blood pressure:** The blood pressure at the peak of each heartbeat.

many people do not know they have it, and others stop taking their blood pressure medications because they dislike the side effects or do not think their drugs are necessary. As of 2008, only 20 percent of Americans diagnosed with high blood pressure were following their doctors' recommendations and getting adequate treatment. Researchers are looking at patient education programs and behavioral approaches to treatment as much as new drugs and new methods of screening for hypertension.

SEE ALSO Diabetes; Heart attack; Heart failure; Obesity; Stroke

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# Hypoglycemia

# Definition

Hypoglycemia is a syndrome (group of related symptoms) caused by abnormally low levels of blood sugar. It is not a disease or disorder by itself. Although some doctors disagree about the exact cutoff point for measuring hypoglycemia, the usual standard is 60–70 milligrams per deciliter (mg/dL) of blood.

# Description

Hypoglycemia can be understood as the result of the body's difficulty in regulating blood sugar levels. It is normal for people's blood sugar levels

Also Known As Low blood sugar, glucose disorder

#### Cause

Difficulty in maintaining a steady level of blood glucose

### Symptoms

Hunger, dizziness, shakiness, feeling anxious or weak, sweating, mental confusion

#### Duration

Minutes to an hour



GlucaGen Hypokit, an emergency treatment for low blood sugar, often seen in diabetics. © MEDICAL-ON-LINE / ALAMY.

to rise and fall over the course of a normal day from about 70 mg/dL to 140 mg/dL, depending on whether they has just eaten, if they are digesting their meal, or if they have not eaten for some hours (as when sleeping). The body normally regulates the level of glucose in the blood by means of two hormones secreted by the pancreas, a small organ located near the liver.

When a person eats a meal, the carbohydrates in such foods as rice, potatoes, pasta, sugary foods, and bread are broken down into glucose, which is then absorbed into the bloodstream. As the glucose level in the blood rises, the pancreas secretes insulin, a hormone that helps the body's tissues make use of the glucose. If there is more glucose in the blood than is needed for the body's energy needs at the time, the extra glucose is stored in the liver in a form called glycogen. As the levels of glucose in the blood drop, the pancreas secretes another hormone called glucagon. Glucagon stimulates the liver to convert the stored glycogen back into glucose and release it into the blood. The additional glucose then raises the person's blood sugar level.

What happens in hypoglycemia is that the normal process of blood sugar regulation no longer works smoothly. This problem may develop as a complication of diabetes, a side effect of some medications, or the result of other diseases or tumors. When blood sugar levels drop below about 70 mg/dL, a person with hypoglycemia may begin to experience the mental and physical symptoms of hypoglycemia.

# Demographics

It is difficult to tell with certainty how many people in the general American population suffer from hypoglycemia, because some people use the term loosely to refer to irritable feelings or mild anxiety associated with hunger even though they have not had a blood sugar test and may in fact have normal levels of blood glucose. Most doctors think that between 5 and 10 percent of Americans have true hypoglycemia. About 55 percent of patients with diabetes will have mild hypoglycemia at some point during treatment for the disease. Hypoglycemia caused by tumors that secrete insulin is very rare, affecting one or two persons per million.

As far as is known, hypoglycemia affects persons of all races and men and women equally. Reactive or fasting hypoglycemia is more common in adults over thirty-five than in adolescents or young adults. Hypoglycemia related to food allergies or overly low levels of growth hormone occurs mostly in children.

#### **Causes and Symptoms**

Hypoglycemia has a number of possible causes: In patients with diabetes, hypoglycemia is usually a side effect of the medications taken to control blood sugar levels. A person's blood sugar level can fall too low if he or she skips meals, exercises too long or too vigorously, takes too large a dose of their diabetes medication, or drinks alcohol.

Patients who do not have diabetes can develop reactive hypoglycemia. This is a condition in which a person's blood sugar drops suddenly between two and five hours after eating sugary foods. Reactive hypoglycemia is not caused by a disease.

Fasting hypoglycemia is a condition that develops in some people as a result of tumors that secrete insulin; certain types of hormone disorders; drinking alcohol; or taking certain medications (particularly sulfa drugs, quinine, and aspirin). It is most noticeable when a person wakes up in the morning.

The symptoms of hypoglycemia are related to the functioning of the central nervous system (CNS) and another part of the nervous system called the sympathetic nervous system. The reason why low blood sugar affects the nervous system before other parts of the body is that the brain and the nerves have higher energy requirements than other tissues. If blood sugar levels drop too low, a hormone called epinephrine is released, which triggers both mental and physical symptoms related to the nervous system.

Mental symptoms that are caused by hypoglycemia include confusion, difficulty thinking clearly, and eventually loss of consciousness or seizures.

Physical symptoms typically include sweating or a clammy feeling, headaches, general weakness or dizziness, speeded-up heartbeat, trembling or shaking, and hunger.

Not everyone with hypoglycemia experiences the same symptoms or experiences them with the same degree of severity. It is possible for a person to have a blood sugar level below 60 mg/dL and have no noticeable symptoms.

#### Diagnosis

Diagnosis of hypoglycemia is based in part on the patient's history and partly on the results of blood and other tests. If the patient does not have diabetes, the doctor will look for three signs known as Whipple's triad: 1) the patient has the symptoms of hypoglycemia; 2) when tested, the blood sugar level is below 45 mg/dL (in a woman) or 55 mg/dL (in a man); 3) the symptoms are relieved in a few minutes when the patient is given sugar or a sugary drink.

If the patient has diabetes, the doctor will review the patient's treatment history to see whether the dosage or specific drug needs to be adjusted. The doctor may also order laboratory tests to look for breakdown products of insulin in the patient's blood. If the person has an insulin-secreting tumor, their blood insulin levels will be high but the level of insulin breakdown products will be low.

Reactive hypoglycemia is diagnosed by measuring the person's blood glucose in the doctor's office while he or she is having symptoms and then measuring the blood glucose again after the patient eats or drinks. If the patient's blood glucose level was below 70 mg/dL while he or she was having symptoms and the symptoms were relieved by food, the person is diagnosed as having reactive hypoglycemia.

Fasting hypoglycemia is diagnosed by a blood sample that shows a blood glucose level of less than 50 mg/dL after an extended supervised fast (usually seventy-two hours in an adult). A healthy person can usually maintain a blood glucose level above 50 mg/dL for seventy-two hours.

# Treatment

Treatment for hypoglycemia depends in part on its cause. Diabetics are usually asked to monitor their lifestyle habits, particularly eating and exercise patterns, as well as paying close attention to the proper use of their medications.

Patients with reactive hypoglycemia are cautioned to avoid sugary foods; have starches, high-protein, and high-fiber foods instead; and eat small meals or snacks every three to four hours rather than three large but widely spaced meals.

Fasting hypoglycemia is usually treated by evaluating the patient's medications and adjusting dosages as necessary; and recommending avoidance of alcohol. Fasting hypoglycemia caused by tumors is treated by surgical removal of the tumor.

# Prognosis

Most people recover completely from an episode of hypoglycemia within minutes of taking some form of glucose. In a few cases, people who have fallen into comas before they were treated suffer long-term brain damage. In a very few cases, people may die from hypoglycemia if not treated.

# Prevention

Preventive measures vary somewhat for diabetics and for nondiabetics with hypoglycemia. Patients with diabetes should take the following steps to prevent hypoglycemia:

- Take medications exactly as prescribed; measure doses carefully.
- Do not skip meals or eat less than the amount of food prescribed for the insulin dosage.
- Keep alcohol consumption to a minimum.
- Exercise in moderation and check the blood glucose level before exercising.
- Check the blood glucose level with a home meter if early symptoms of hypoglycemia appear. If the level is below 70 mg/dL, take some glucose in the form of five or six pieces of hard candy, 1–2 teaspoons of honey or table sugar, two or three glucose tablets, or one-half cup of fruit juice.
- Measure the blood sugar again in fifteen minutes. If it is still below 70, take another dose of sugar or a sugary food or beverage. Always carry one of these foods or drinks in case of need.

# WORDS TO KNOW

- **Fasting hypoglycemia:** A type of hypoglycemia in people without diabetes that is caused by hormone deficiencies, medication side effects, or tumors rather than by reaction to a sugar-rich meal.
- **Glucagon:** A hormone secreted by the pancreas that raises blood sugar levels by signaling the liver to convert glycogen to glucose.
- **Glycogen:** A form of glucose that is stored in the liver as an energy reserve.

**Insulin:** A hormone secreted by the pancreas that lowers blood sugar levels by allowing

body tissues to absorb and make use of the glucose.

- **Reactive hypoglycemia:** A condition in which a person develops hypoglycemia between two and five hours after eating foods containing high levels of glucose.
- Whipple triad: A group of three factors used to diagnose hypoglycemia: symptoms; blood sugar measuring below 45 mg/dL for a woman and 55 mg/dL for a man; and rapid recovery following a dose of sugar.
- Wear a medical identification tag or bracelet if you have ever lost consciousness as a result of hypoglycemia.
- Ask the doctor about having a glucagon kit at home or work. People with a history of severe hypoglycemia may need to have a friend or relative inject them with the glucagon if they lose consciousness.
- Never drive a car without checking to see that the blood glucose level is above 70 mg/dL.
- People with reactive hypoglycemia should consult a registered dietitian to help them plan a personalized diet that will lower their risk of hypoglycemic episodes while still allowing them to eat foods they enjoy.
- Follow-up visits to the doctor to evaluate any further symptoms are also an important part of preventive care.

# The Future

Research is focusing on a better understanding of the causes of reactive hypoglycemia. Researchers are also studying whether new devices for monitoring blood glucose levels frequently at home will help reduce the risk of hypoglycemic episodes.

### SEE ALSO Diabetes

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Also Known As

Core temperature drop

#### Cause

Exposure to cold; surgical procedure; diseases related to aging

#### Symptoms

Intense shivering; loss of muscle and mental control; heart failure and death

#### Duration

Depends on length of cold exposure and overall health



# Hypothermia

# Definition

Hypothermia is defined as a subnormal body temperature caused by the loss of more body heat than the body can replace. There are several different types of hypothermia. Accidental hypothermia is caused by exposure to cold weather, while intentional or induced hypothermia is a medical technique used to increase a patient's chances of recovery after stroke or cardiac arrest. Primary hypothermia is caused by exposure to a cold or wet environment; secondary hypothermia refers to lowering

#### Hypothermia

Images of body heat on a warm day (left) and a cold day (right). Red and pink are warmer areas; blue areas are colder. CLAUS LUNAU / BONNIER PUBLICATIONS / PHOTO RESEARCHERS, INC



of body temperature caused by such diseases as Parkinson disease, multiple sclerosis, or brain tumors. Many elderly people with hypothermia have the secondary form.

# Description

The normal temperature of the human body is about 97.9°F (36.6°C). Body temperature is regulated by a part of the brain called the hypothalamus. When a person begins to feel cold, the hypothalamus releases chemicals to step up heat production from the body's energy stores. The muscles begin to shiver, which also releases heat. Shivering increases the body's rate of heat production by two to five times.

The hypothalamus also works to control heat loss by slowing down the flow of blood to the arms, hands, legs, and feet. This process is known as vasoconstriction. Vasoconstriction protects the body by helping to keep the heart and other vital organs in the body core functioning as effectively as possible. In most cases, the person will feel uncomfortable enough to go back indoors or put on more clothing. If the person cannot find shelter or otherwise get warm, the person's central nervous system

# An Evil Experiment

In the early 1940s Nazi doctors conducted cruel experiments on Russian prisoners of war and concentration camp inmates to see how long it takes people to freeze to death and to try various ways to rewarm people with hypothermia. The reason for these experiments was the high casualty rate suffered by German soldiers during the winter of 1941–1942 following Hitler's invasion of the Soviet Union.

The men used for these experiments were exposed to cold either by being put in vats of icy water or tied naked to stretchers and placed outside in freezing weather. The prisoners put in cold water had probes inserted into their rectums that measured the drop in their body temperature; it was found that they died when their core temperature dropped to 77°F (25°C).

The methods used to warm the men were just as cruel: using sun lamps so hot they burned the skin and forcing scalding-hot water into the stomach, intestines, or bladder. None of the prisoners survived these rewarming treatments.

One of the doctors who conducted these experiments, Sigmund Rascher, was shot on orders of the Nazi leaders in April 1945 before the Allies liberated the concentration camps. Some doctors involved in the experiments were tried for war crimes in 1947 and either executed or sentenced to long prison terms. (CNS) will eventually be affected and the hypothalamus will no longer be able to regulate the body's temperature. When temperature regulation fails, the heart and other vital organs can no longer function properly.

Hypothermia may come on gradually; the person may not know that his or her body temperature is dropping, though others may notice the person is becoming irritable, slurring their speech, becoming clumsy in their movements, or showing poor judgment. The rate at which a person develops hypothermia depends on basic health, environmental temperature and humidity, and the warmth and dryness of their clothing.

Doctors distinguish three stages of hypothermia according to severity:

- Mild hypothermia: body temperature between 95–90°F (35–32°C).
- Moderate hypothermia: body temperature between 90–82°F (32–28°C).
- Severe hypothermia: body temperature below 82°F (28°C).

# **Demographics**

The true number of people who develop hypothermia in the United States each year is not known; doctors think that those who are taken to hospital emergency rooms are only a small fraction of the actual number of cases. According to the Centers for Disease Control

and Prevention, between 650 and 700 people die from hypothermia each year in the United States, or about 0.2 persons per 100,000. Cold-related deaths are twice as common in men as in women. Fifty-two percent of the victims were over the age of sixty-five. The states with the highest rates of deaths from hypothermia are Alaska, New Mexico, North Dakota, Montana, North and South Carolina, and Arizona. Some states have higher-than-average rates because they experience rapid temperature changes at certain times of the year or have mountainous regions with rapid changes in overnight temperatures.

Some people are at greater risk of hypothermia, particularly infants and young children and the elderly. People with diabetes and other illnesses that affect blood circulation are also more likely to develop hypothermia when exposed to cold.

### **Causes and Symptoms**

The most common causes of hypothermia are loss of body heat due to exposure to cold weather or to a combination of cold and dampness. Swimmers or people who fall into a body of water can develop hypothermia even when the water is not icy cold. According to the U.S. Coast Guard, a person who falls into water at 40°F (4°C) will become unconscious in about half an hour and die within another hour; at 32°F (0°C), survival time drops to fifteen minutes.

The symptoms of hypothermia include mental as well as physical symptoms:

- Mild hypothermia: The person shivers vigorously and begins to show confusion and poor judgment. They become moody, may slur their speech, move clumsily, and start breathing heavily.
- Moderate hypothermia: The person's breathing slows down, central nervous activity is lowered and the body loses its ability to generate additional heat by shivering. The heart may develop an abnormal rhythm. A symptom sometimes seen at this stage is paradoxical undressing, in which the person becomes confused, disoriented, and starts to remove their clothing—which speeds up the rate of heat loss. Between 20 and 50 percent of deaths from hypothermia are thought to result from paradoxical undressing.
- Severe hypothermia: The risk of heart failure increases; the person may go into coma and be unresponsive when touched. There may be no pulse and the blood pressure may be abnormally low. The lungs may fill with fluid and breathing may be difficult.

### Diagnosis

Diagnosis of hypothermia is usually based on a measurement of body temperature in the emergency room, as many of the physical and mental symptoms of hypothermia can be caused by such other conditions as stroke, alcohol intoxication, medication side effects (common in the elderly), and mental illness. The use of a special low-temperature probe inserted into the rectum or the bladder is thought to give a more accurate reading than a standard thermometer.

# Treatment

The first line of emergency treatment for hypothermia includes preventing further heat loss, raising the body core temperature, and preventing heart failure. To lower the risk of the person's developing an abnormal heart rhythm, rescuers are advised to move the person as gently as possible and to begin rewarming the person in the field before taking him or her to the hospital. Wet clothing is removed and replaced with dry clothing and blankets or a dry sleeping bag. Hot water bottles or chemical heat packs are used to warm the body; in extreme emergencies, rescuers can warm the person by skin-to-skin contact.

In the hospital, emergency room doctors will take the person's core temperature to determine the severity of the hypothermia; the lower the body temperature, the more careful the doctors must be in rewarming the patient. Sometimes the person's core temperature continues to drop after rewarming is started; this complication, known as after drop, is thought to result from cooler blood in the patient's extremities being recirculated back into the body's core organs during rewarming.

Various techniques have been used to rewarm people with hypothermia, ranging from wrapping the patient in heated blankets, immersing him or her in warm water in a device known as a Hubbard tank, or giving warmed and humidified oxygen through a face mask or endotracheal tube. Another method that is used is injection of intravenous fluids heated to  $113^{\circ}$ F (45°C).

### Prognosis

The prognosis for recovery from hypothermia depends on its severity. Most people survive mild hypothermia without significant after-effects. The death rate for moderate hypothermia is close to 21 percent; for severe hypothermia, it is close to 40 percent.

Hypothermia can lead to a number of long-term health complications, including frostbite, pneumonia, other infections, disorders of the pancreas and bladder, and lung damage.

# Prevention

Accidental hypothermia can be prevented by dressing sensibly for coldweather activities and by avoiding the use of alcohol and other substances that interfere with good judgment. In the summer, people should be careful not to drink before operating a boat, as falling into water even in summertime temperatures can still cause hypothermia.

In addition, persistent shivering is a sign that the body is losing too much heat; this is an important signal to go back inside as soon as possible. A tip that can help people remember how to dress for winter is the word COLD:

- Cover: Keep head, neck, and face covered with a warm hat, hood, or scarf. Mittens are better than gloves for protecting the hands because they keep the fingers closer together.
- Overexertion: Avoid exercise or other activities that cause heavy sweating, because the combination of moisture on the skin, clothing that becomes damp from sweat, and the cold outside can cause rapid loss of body heat.
- Layer: Loose-fitting layered clothing holds in body heat better than tight-fitting garments. Water-repellent outerwear is a good choice for wet and windy weather.
- Dry: People should stay as dry as possible outdoors, and check mittens and boots from time to time to make sure that snow cannot get inside and melt. It is a good idea to pack an extra pair of dry socks and mittens just in case.

The CDC recommends carrying emergency supplies of food (granola and crackers are good choices), blankets, matches and candles, and extra clothing in the car during the winter in case the car stalls or is stranded in snow. Additional safety precautions include checking the weather forecast before setting out on a trip and letting others know the expected arrival time.

# The Future

Hypothermia is thought to be a growing problem in North America because of the increasing numbers of people participating in outdoor sports as well as the increasing numbers of mentally ill and substanceaddicted homeless people. Public health doctors and social workers often have trouble convincing homeless people to go into public shelters in the

# WORDS TO KNOW

**After drop:** A term that doctors use to refer to lowering of the body's core temperature that continues while the person is being rewarmed.

**Hypothalamus:** The part of the brain that controls body temperature, hunger, thirst, and response to stress.

**Paradoxical undressing:** A symptom sometimes seen in people with moderate or severe

hypothermia, thought to be caused by a malfunction of the hypothalamus. The person becomes confused, disoriented, and begins to remove clothing.

Vasoconstriction: A narrowing of the blood vessels in response to cold or certain medications.

cold weather, however, because they claim they are afraid of drug abusers who also use the shelters.

**SEE ALSO** Frostbite

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# **m** Hypothyroidism

# Definition

Hypothyroidism is a condition in which a person's thyroid gland is not producing enough hormone. It may be caused by an autoimmune disorder, a genetic defect in a newborn, certain medications, surgical removal of the thyroid gland, radiation therapy for cancer, and other reasons. Hypothyroidism is sometimes categorized as either primary (caused by a problem in the thyroid gland itself) or secondary (caused by the lack of hormones that ordinarily stimulate the thyroid gland to produce thyroid hormone).

# Description

Hypothyroidism is an endocrine disorder. It is caused by underfunctioning of a gland that is part of the endocrine system—a group of small organs located throughout the body that regulate growth, metabolism, tissue function, and emotional mood. The thyroid gland itself is a butterfly-shaped organ that lies at the base of the throat below the Adam's apple.

Hypothyroidism is not easy to diagnose because its symptoms are found in a number of other diseases; it often comes on slowly; and it may produce few or no symptoms in younger adults. In general, hypothyroidism is characterized by a slowing down of both physical and mental activities.

# Demographics

About 3 percent of the general population in the United States and Canada have some form of hypothyroidism. Apart from cretinism, which affects one child in every 3,000 to 4,000, hypothyroidism is largely a disease of adults. The most common form of primary hypothyroidism in North America is Hashimoto disease, an autoimmune disorder that is diagnosed in about fourteen women out of every 1,000 and one man in every 2,000. Internationally, however, the most common cause of hypothyroidism is a lack of iodine in the diet. Also Known As Myxedema, if the hypothyroidism is severe

#### Cause

Underproduction of the thyroid hormone

#### Symptoms

Fatigue, weight gain, brittle nails and hair, menstrual problems, sensitivity to cold

#### Duration

Develops slowly over months to years



Woman with hypothyroidism, shown by the swelling of her neck, called a goiter. DR. P. Some people are at increased risk of hypothyroidism:

- Women. Women are two to eight times as likely to have hypothyroidism, depending on the age group being studied.
- Age over fifty. In one Massachusetts study, 6 percent of women over age sixty and 2.5 percent of men over age sixty were found to be hypothyroid.
- Race. According to the National Institutes of Health (NIH), the rates of hypothyroidism in the United States are highest among Caucasians (5.1 percent) and Hispanics (4.1 percent) and lowest among African Americans (1.7 percent).
- Obesity.
- People who have close relatives with an autoimmune disease.

# **Causes and Symptoms**

The most common causes of hypothyroidism are:

- Hashimoto disease. This is an autoimmune disorder in which the patient's immune system attacks the thyroid gland, leading to tissue destruction.
- Treatment for hyperthyroidism. People who have been treated for an oversupply of thyroid hormone with radioactive iodine may lose their ability to produce enough thyroid hormone.
- Surgery on the thyroid gland.
- Radiation therapy for the treatment of head or neck cancer.
- Medications. Lithium, given to treat some psychiatric disorders, and certain heart medications may affect the functioning of the thyroid gland.
- Pregnancy. As many as 10 percent of women may become hypothyroid in the first year after childbirth, particularly if they have diabetes.

# An Olympic Champion's Story

Carl Lewis (1961–), a track and field superstar, was the first athlete to equal Jesse Owens's (1913– 1980) feat of winning four gold medals in a single Olympic Games in 1936. In 1984 Lewis won his four medals at the Games held in Los Angeles. He continued to dominate track and field events into the 1990s.

Shortly before competing in the 1996 Olympic Games in Atlanta, however, Lewis was diagnosed with hypothyroidism. His early symptoms were easy to miss, such as a weight gain that could be explained as the result of a muscle-building program that was part of his training for the games. He began treatment with synthetic thyroid hormone at once. In July 1996, he won the long jump at the Atlanta Games and became one of only several athletes in the history of the Olympics to win a total of nine gold medals in the course of his career.

Lewis later published a book about his condition in which he said, "As is the case with most people with thyroid conditions, I had no clue that I had the condition at all. The fact that I was checked was a fortunate accident.... I knew nothing about thyroid problems before discovering that I had one



Carl Lewis in the 1992 Olympics. AP IMAGES.

myself. Educate yourself, and follow prescribed treatment. As I showed in the Olympics, you can go back to being 100 percent. I feel even better now than I did in Atlanta, now that my stress level is on a more even keel and my medication levels are just right."

- Viral infections. These can cause a short-term inflammation of the thyroid gland in some people.
- A tumor in the pituitary gland. The pituitary gland produces a hormone called thyroid-stimulating hormone or TSH. Low levels of TSH can lead to secondary hypothyroidism.
- Congenital. About one baby in every 3,000 to 4,000 is born with a defective thyroid gland or no gland at all.
- Too little iodine in the diet. This cause of hypothyroidism is most common in developing countries; it is rare in North America and Europe.

Hypothyroidism can be difficult to diagnose because many of its early symptoms are not unique to it. In addition, the symptoms typically come on gradually. The person may simply feel tired or less energetic than usual, or develop dry, itchy skin and brittle hair that falls out easily. The classic symptoms of hypothyroidism—sensitivity to cold, puffy complexion, decreased sweating, and coarse skin—may occur in only 60 percent of patients. It may take months to years before the person or his or her doctor begins to suspect a problem with the thyroid gland.

Not every patient with an underactive thyroid has the same symptoms or has them with the same severity. Common symptoms of hypothyroidism, however, include the following:

- · Increased sensitivity to cold weather
- Dry, itchy skin and a pale or yellowish complexion
- Dry brittle hair that falls out easily and nails that break or split
- Constipation
- Goiter (swelling in the front of the neck caused by thyroid enlargement)
- Hoarse voice and puffy facial skin
- Unexplained weight gain of 10–20 pounds (4.5–9 kilograms), most of which is fluid
- Sore and aching muscles, most commonly in the shoulders and hips
- In women, extra-long menstrual periods or unusually heavy bleeding
- Weak leg muscles
- · Decreased sweating
- Arthritis
- Memory loss or difficulty concentrating
- Slowed heart rate (less than 60 beats per minute) and lowered blood pressure
- Depression

### Diagnosis

The diagnosis of hypothyroidism is usually made by tests of the patient's thyroid function following a careful history of the patient's symptoms. The first test is a blood test for thyroid-stimulating hormone, or TSH.

TSH is a hormone produced by the pituitary gland in the brain that stimulates the thyroid gland to produce thyroid hormone. When the thyroid gland is not producing enough hormone, the pituitary gland secretes more TSH; thus a high level of TSH in the blood indicates that the thyroid gland is not as active as it should be.

The TSH test, however, does not always detect borderline cases of hypothyroidism. The doctor may order additional tests to measure the levels of thyroid hormone as well as TSH in the patient's blood. If the doctor thinks that the patient may have Hashimoto disease, he or she may test for the presence of abnormal antibodies in the blood. Because Hashimoto disease is an autoimmune disorder, there will be two or three types of anti-thyroid antibodies in the patient's blood in about 90 percent of cases.

In some cases, the doctor may also order an ultrasound study of the patient's neck in order to evaluate the size of the thyroid gland or take a small sample of thyroid tissue in order to make sure that the gland is not cancerous.

### Treatment

Treatment for hypothyroidism consists of a daily dose of a synthetic form of thyroid hormone sold under the trade names of Synthroid, Levothroid, or Levoxyl. The patient is told that the drug must be taken as directed for the rest of his or her life.

In the early weeks of treatment, the patient will need to see the doctor every four to six weeks to have his or her TSH level checked and the dose of medication adjusted. After the doctor is satisfied with the dosage level and the patient's overall health, checkups are done every six to twelve months. The reason for this careful measurement of the medication is that too much of the synthetic hormone increases the risk of osteoporosis in later life or abnormal heart rhythms in the present.

Congenital hypothyroidism or cretinism is also treated with synthetic thyroid hormone. Most hospitals now screen newborns for thyroid problems, because untreated hypothyroidism can lead to lifelong physical and mental developmental disorders.

# Prognosis

The prognosis for patients with hypothyroidism is very good, provided they take their medication as directed. They can usually live a normal life with a normal life expectancy. Children with congenital hypothyroidism have a good prognosis if the disorder is caught and treated early. Some develop learning disorders, however, in spite of early treatment.

The chief risks to health are related to a lack of treatment for hypothyroidism. If low levels of thyroid hormone are not diagnosed and treated, patients are at increased risk of goiter, an enlarged heart, and severe depression. In addition, women with untreated hypothyroidism have a higher risk of giving birth to babies with cleft palate and other birth defects.

One rare but potentially life-threatening complication of long-term untreated hypothyroidism is myxedema coma. In this condition, which is usually triggered by stress or illness, the person becomes extremely sensitive to cold, may be unusually drowsy, or lose consciousness. Heart rate, blood pressure, and breathing may all be abnormally low. Myxedema coma requires emergency treatment in a hospital with intravenous thyroid hormone and intensive care nursing.

### Prevention

There are no proven ways to prevent hypothyroidism because the disorder has so many possible causes.

### The Future

Research in hypothyroidism in the early 2000s has a number of different goals. One is to look for specific genes that may be linked to hypothyroidism. Another area of research is to discover reasons for the high female/male ratio. Still another goal is to discover a cure for the condition that will do away with the need for lifetime treatment with synthetic thyroid hormone.

SEE ALSO Hashimoto disease

### For more information

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# WORDS TO KNOW

**Congenital:** Present at birth.

**Cretinism:** A form of hypothyroidism found in some newborns.

- **Endocrine system:** A system of small organs located throughout the body that regulate metabolism, growth and puberty, tissue function, and mood. The thyroid gland is part of the endocrine system.
- **Goiter:** A swelling in the neck caused by an enlarged thyroid gland.
- **Hyperthyroidism:** A disease condition in which the thyroid gland produces too much thyroid hormone.

- **Hypothyroidism:** A disease condition in which the thyroid gland does not produce enough thyroid hormone.
- **Metabolism:** The chemical changes in living cells in which new materials are taken in and energy is provided for vital processes.
- **Myxedema:** A synonym for hypothyroidism. Myxedema coma is a condition in which a person with untreated hypothyroidism loses consciousness. It is potentially fatal.

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Genetic Infection

Injury

Multiple

Other

Unknown



# Infectious Diseases

Infectious diseases are a group of illnesses caused by various microbes (organisms too small to be seen without a microscope), which may be bacteria, viruses, fungi, protozoa, parasites, or abnormal proteins called prions. Infectious diseases are contagious. They may spread by physical contact with other infected persons or contaminated objects; by contact with body fluids; by eating or drinking contaminated food or water; by airborne droplets; or by vectors—insects or other animals that transmit disease organisms to humans.

Infectious diseases can spread over a wide geographical area, causing epidemics or pandemics. Some historical pandemics that caused widespread loss of life include the Black Death in the fourteenth century; the smallpox epidemics in Europe and the Americas in the eighteenth century; and the influenza pandemic of 1918–1919.

The microbes that cause infectious diseases are defined as either primary or opportunistic depending on whether they cause disease in people with normal immune systems or whether they can cause disease only in people with weakened immune systems, also called opportunistic infections.

Infectious diseases are still a major cause of death worldwide. According to the World Health Organization (WHO), about 15 million people die each year of infectious diseases—about 26 percent of all deaths. The major killers as of 2008 were pneumonia and influenza, diarrheal diseases, AIDS, and malaria.

**SEE ALSO** AIDS; Anthrax; Avian influenza; Bronchitis; Chickenpox; Chlamydia; Cold sore; Common cold; Conjunctivitis; Creutzfeldt-Jakob disease; Ear infection; Ebola and Marburg hemorrhagic fevers; Encephalitis; Food poisoning; Genital herpes; Gonorrhea; Hantavirus infection; Hepatitis A; Hepatitis B; Hepatitis C; HPV infection; Infectious mononucleosis; Influenza; Lyme disease; Malaria; Measles; Meningitis; Necrotizing fasciitis; Periodontal disease; Plague; Pneumonia; Polio; Rabies; Rheumatic fever; Rubella; Scarlet fever; Severe acute respiratory syndrome; Smallpox; Sore throat; Staph infection; Strep throat; Syphilis; Tetanus; Tooth decay; Toxic shock syndrome; Toxoplasmosis; Tuberculosis; Ulcers; Urinary tract infection; Warts; West Nile virus infection; Whooping cough



# Infectious Mononucleosis

# Definition

Infectious mononucleosis is a highly contagious disease caused by the Epstein-Barr virus (EBV) and spread primarily by contact with the saliva of an infected person. Although the disease is sometimes known as the kissing disease because of the role of saliva in spreading the infection, mononucleosis can also be spread through blood and genital secretions (although these forms of transmission are very rare).

# Description

EBV is a very common virus worldwide; most people become infected with it at some point in their lives. Many people become infected with the virus and never develop noticeable symptoms. Those who do develop symptoms typically experience about two weeks of fever, sore throat, and swollen lymph nodes in the neck, throat, or armpits. Although mononucleosis is not a major threat to health, it is a common cause of absence from school or work in teenagers and young adults because it can lead to weeks or months of fatigue and lowered energy.

#### Also Known As

Mono, glandular fever, kissing disease

# Cause

Epstein-Barr virus (EBV)

#### Symptoms

Fever, sore throat, swollen lymph nodes

#### Duration

Usually two to six weeks; sometimes several months

#### Infectious Mononucleosis



White areas at the back of the throat of a patient with infectious mononucleosis. © MEDICAL-ON-LINE/ALAMY.

### **Demographics**

In the United States, mononucleosis is most common in teenagers and young adults; it is more common in younger children in developing countries. People in any age group can get the disease if they are exposed, however. As many as 95 percent of American adults between thirty-five and forty years of age have been infected, although not all of these have had the symptoms of the illness. When infection with EBV occurs during adolescence or young adulthood, it causes infectious mononucleosis between 35 and 50 percent of the time.

Males and females are equally likely to get mononucleosis, as are people of all races and ethnic groups.

# **Causes and Symptoms**

Infectious mononucleosis is caused by the Epstein-Barr virus, or EBV. The virus is normally transmitted by contact with the saliva of an infected person; it is not ordinarily transmitted through the air. The virus takes about four to six weeks to incubate, and thus infected persons can spread the disease to others over a period of several weeks. After entering the patient's mouth and upper throat, the virus infects B cells, which are a certain type of white blood cell produced in the bone marrow. The infected B cells are then carried into the lymphatic system, where they affect the liver and spleen and cause the lymph nodes to swell and

enlarge. The infected B cells are also responsible for the fever, swelling of the tonsils, and sore throat that characterize mononucleosis.

After the symptoms of mononucleosis go away, the EBV virus remains in a few cells in the patient's throat tissues or blood for the rest of the person's life. The virus occasionally reactivates and may appear in samples of the person's saliva, it does not cause new symptoms of illness, it may be transmitted (given) to a susceptible person. Mononucleosis does not cause any problems during pregnancy, such as miscarriages or birth defects.

The primary symptoms of mononucleosis are fever, sore throat, and swollen lymph nodes in the throat, armpit, or neck. Other common symptoms include:

- Swelling or enlargement of the spleen or liver
- General discomfort and mild muscle aches
- Sleepiness and fatigue
- Loss of appetite
- Skin rash
- · Swollen tonsils or a yellowish coating on the tonsils
- Night sweats

Less common symptoms of mononucleosis that some patients experience include:

- Headache
- Stiff neck
- Sensitivity to light
- Shortness of breath and chest pain
- Nosebleed
- Hives
- Jaundice

# Diagnosis

The diagnosis of mononucleosis is usually based on the results of blood tests combined with the doctor's examination of the patient's throat and neck. The doctor will also tap on or feel the patient's abdomen to see whether the liver and spleen have become enlarged.

A patient infected by EBV will have an increased number of certain white blood cells in the blood sample called atypical lymphocytes, and antibodies to the Epstein-Barr virus. These antibodies can be detected by a test called the monospot test, which gives results within a day but may not be accurate during the first week of the patient's illness. Another type of blood test for EBV antibodies takes longer to perform but gives more accurate results within the first week of symptoms.

### Treatment

There is no cure for mononucleosis because it is caused by a virus; it cannot be treated by antibiotics. Treatment consists of self-care at home until the symptoms go away. Patients should rest in bed if possible and drink plenty of fluids. Non-aspirin pain relievers like Advil or Tylenol can be taken to bring down the fever and relieve muscle aches and pains. Throat lozenges or gargling with warm salt water may help ease the discomfort of a sore throat.

Because mononucleosis can affect the spleen, patients should avoid vigorous exercise or contact sports for at least one month after the onset of symptoms or until the spleen returns to its normal size. This precaution will lower the risk of rupture of the spleen.

#### Prognosis

Mononucleosis rarely causes serious complications. In most patients, the fever goes down in about ten days, but fatigue may last for several weeks or months. Some people do not feel normal again for about three months. A patient who feels sick longer than four months, however, should go back to the doctor to see whether they have another disease or disorder in addition to mononucleosis. In some cases, the patient is diagnosed with chronic fatigue syndrome or CFS. The Epstein-Barr virus does not cause CFS; however, it appears to make some patients with mononucleosis more susceptible to developing chronic fatigue syndrome.

One way to speed complete recovery from infectious mononucleosis is to get plenty of rest early in the disease; the more rest patients get at the beginning, the more quickly they recover.

Severe complications of mononucleosis are unusual but may include rupture of the spleen, which occurs in 0.5 percent of patients—almost all of them males who returned to sports too quickly. Airway obstruction may develop in one patient per thousand, most often a small child. This complication can be treated with steroid medications. Between 1 and 3 percent of patients may develop a form of anemia that can also be treated with steroids.

# WORDS TO KNOW

- Anemia: A condition in which there are not enough red cells in the blood or enough hemoglobin in the red blood cells.
- **B cell:** A type of white blood cell produced in the bone marrow that makes antibodies against viruses.
- Jaundice: A yellowish discoloration of the skin and whites of the eyes caused by increased levels of bile pigments from the liver in the patient's blood.

#### Prevention

There is no vaccine that can prevent mononucleosis. In addition, the fact that many people can be infected with the virus and transmit it to others without having symptoms of the disease means that mononucleosis is almost impossible to prevent. The best precautionary measure is for patients who have been diagnosed with mono to avoid kissing, or other close personal contact with, others and to wash their drinking glasses, food dishes, and eating utensils separately from those of other family members or friends for several days after the fever goes down. It is not necessary for people with mono to be completely isolated from other people, however.

Because the Epstein-Barr virus remains in the body after the symptoms of mononucleosis go away, people who have had the disease should not donate blood for at least six months after their symptoms started.

# **The Future**

As of 2008 researchers were working on a vaccine against EBV. In December 2007 the *Journal of Infectious Diseases* reported that a vaccine developed in Belgium shows promise in preventing mononucleosis. The vaccine must undergo further clinical trials, however, before it can be definitely shown to be effective and licensed for use in the United States.

**SEE ALSO** Chronic fatigue syndrome

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# Influenza

# Definition

Influenza (flu) is a highly infectious disease of mammals and birds caused by a family of viruses. There are three basic types of influenza virus, known as A, B, and C. Most cases of flu in humans are caused by influenza A.

# Description

Influenza is an illness of the nose and throat that has a long history as a troublemaker. It is one of the most highly infectious diseases that affect humans, being the cause of numerous pandemics (large-scale outbreaks), some of which have spread around the civilized world. The first influenza pandemic that is known to have been global in scale took place in 1580; it started in China and spread across Central Asia to Africa and then to Europe, where it nearly wiped out the populations of several major cities in southern Italy and Spain.

While mild cases of influenza are easy to confuse with the common cold, most people will start to feel much sicker with the flu within a few hours of the onset of symptoms. It is not unusual for people with flu to be able to tell the doctor exactly when they first started to feel sick. The Also Known As Flu, Spanish fever

**Cause** Virus

#### Symptoms

Chills, fever, headache, muscle aches and pains, sore throat, coughing

**Duration** One to two weeks

#### Influenza

Viruses reproduce by infecting a body cell and forcing the cell to grow more viruses. Here, the new viruses are emerging from the infected cell. © SPL/PHOTO RESEARCHERS, INC.



first sign is usually fever and chills, followed by a sore throat, pain in the muscles, and feeling extremely tired and weak. Some people have a runny nose. Coughing is usually a later symptom.

Most people who get influenza feel better in one or two weeks; however, the disease can cause serious complications because it makes it easier for people to get other infections, such as bacterial pneumonia and ear infections. It also makes chronic health problems like asthma, chronic bronchitis, emphysema, and congestive heart failure worse. The World Health Organization (WHO) estimates that about 300,000 people around the world die each year from complications of influenza. It is important to note that some diseases that are commonly called the flu (like the stomach flu) are not caused by the influenza virus.

# **Demographics**

Millions of people around the world are infected with influenza each year, although the number and the severity of the reported cases vary from year to year, depending on the severity of the particular strain of virus involved. For example, there were more serious cases of flu reported in the United States in the winters of 1999–2000 and 2003–2004 than in the winters of 2001, 2002, and 2005.

# The Pandemic of 1918–1919

The influenza pandemic of 1918–1919 is one of the deadliest pandemics in world history. It is estimated that between 2 and 5 percent of the world's population died before the disease ran its course. In the first 25 weeks of the pandemic, 25 million people died—as many as died during the first 25 years of the AIDS pandemic.

An outbreak of influenza at Fort Riley in Kansas in March 1918 is generally considered the beginning of the pandemic. Since 200,000 American soldiers were sent to Europe in late March and April of 1918, some of them carried the deadly virus with them. The virus returned to the United States with a vengeance in the summer and fall of 1918, as well as spreading beyond Europe and North America to Asia and Africa. It was unusual not only for its high death rate—about 2–3 percent of those infected, compared to a usual mortality rate of 0.1 percent (or less)—but also for its demographics. In most flu epidemics, the very young and the very old are most at risk for complications. The pandemic of 1918–1919, however, disproportionately affected young adults; more than half of all deaths were reported in people between the ages of 20 and 40.

The other unusual feature of this particular pandemic was the severity of the symptoms and the rapidity of death. This flu caused people to bleed from the nose, stomach, and intestines. Doctors performing autopsies on flu victims were shocked to find the lungs filled with bloody, foamy fluid. People sometimes collapsed on the street and died within hours. A nurse at Hartford Hospital in Connecticut recounted an incident that was typical of the pandemic. Four Yale students had gotten off the train in Hartford because they did not feel well and walked from the train station to the hospital. By the next morning all four young men were dead.



An emergency hospital near Fort Riley, Kansas, during the 1918 influenza pandemic. AP IMAGES.

Flu epidemics usually occur in the winter in temperate climates; however, they can occur at any time of year in countries with tropical climates. Pandemics typically occur every ten to twenty years.

Influenza is most likely to lead to serious complications in infants, the elderly, smokers, people with asthma, people with weakened immune systems, and women in the last three months of pregnancy. The disease affects people of both sexes and all races equally, as far as is known.

#### **Causes and Symptoms**

Most cases of flu in humans are caused by either influenza A or B viruses. The influenza A viruses are most likely to cause pandemics in the human population because they infect wild birds and domestic poultry like chickens and turkeys and thus can easily be transmitted to farmers, hunters, and others who raise or study birds. These viruses can also infect horses, dogs, pigs, camels, cats, ferrets, harbor seals, and whales. In addition, influenza A viruses cause more severe disease; viruses of this type are known to have caused the great pandemic of 1918–1919, the Asian flu epidemic of 1957, the Hong Kong flu epidemic of 1968, and the avian (bird) flu of 2007. Influenza viruses change quickly over time, and flu vaccines must be reformulated every year because of this rapid rate of mutation.

Flu is spread by inhaling droplets from an infected person's coughing or sneezing that contain the virus. It can also be transmitted by kissing or by touching food utensils, handkerchiefs, telephone receivers, doorknobs, desk surfaces, and other objects that may have been handled by an infected person. People can transmit the virus for about twenty-four hours before they start to feel sick, which is another reason why the disease spreads so rapidly during flu season. Adults are infectious for about seven days after they feel sick, and children are infectious for about ten days.

When the virus enters the body, it attaches itself to the moist tissues lining the nose, throat, and lungs, where it invades the cells of these tissues and multiplies rapidly.

Typical flu symptoms include:

- Sudden onset of fever and chills (The fever may range from 100°F [37.8°C] to as high as 104°F [40°C].)
- Flushed face and red watery eyes
- Muscle pains and cramps, which can be quite severe in some people

- Headache
- Dry cough and sore throat
- Runny nose
- Weakness and severe fatigue, often requiring complete bed rest

# Diagnosis

Most people are likely to consult a doctor when their cough becomes troublesome. In most cases the doctor will make the diagnosis of flu on the basis of the patient's symptoms, because the available medical tests are either too slow for the diagnosis to benefit the patient or not costeffective. The definitive diagnosis of flu is based on growing samples of the patient's nose or throat secretions in a laboratory; this process takes three to seven days. Rapid diagnostic tests of blood samples require trained laboratory technicians and equipment.

In 2004 the Food and Drug Administration (FDA) approved a new rapid test for flu called the QuickVue test. The test can be used at the patient's bedside. The doctor swabs the inside of the nose with an applicator that is then soaked in a chemical solution for a minute to extract the virus. A special strip is then inserted into the solution and allowed to remain for about ten minutes. If the flu virus is present, a red or pink line will appear on the strip. The QuickVue test is about 80 percent accurate.

# Treatment

Most people can care for themselves at home by staying away from others, resting in bed, drinking plenty of fluids, and avoiding the use of alcohol and tobacco. Orange juice or sports drinks are better than plain water for preventing dehydration because they contain electrolytes, which are minerals that the body loses during a high fever along with water from body tissues. Fever and muscle aches and pains should be treated with acetaminophen (Tylenol), ibuprofen (Advil or Motrin), or naproxen (Aleve); aspirin should not be used to bring down fever because of the small risk of Reye syndrome in children. Nasal decongestants can be used to clear a runny or stuffy nose if needed.

Some drugs known as antiviral medications can be given to shorten a flu attack or decrease its severity. Two of these, however, are no longer commonly recommended because the type of influenza A virus most active in the United States since 2005 has developed resistance to these medications. They are Symmetrel and Flumadine. People who need an antiviral medication should ask their doctor for either Relenza or Tamiflu, because the flu virus has not yet developed resistance to these drugs. Antiviral drugs must generally be taken within 48 hours after the start of symptoms in order to be effective, however; they also produce such side effects as nausea and vomiting.

#### Prognosis

Most people recover from influenza, although some may feel weak and tire easily for several weeks after the acute symptoms go away. People who develop bacterial pneumonia as a complication of influenza, however, are at some risk of death; the average mortality rate for a flu epidemic is about 0.1 percent. The Centers for Disease Control and Prevention (CDC) estimates that between 20,000 and 30,000 people die each year in the United States as a result of influenza.

### Prevention

People can lower their risk of getting flu by such common-sense precautions as washing hands with soap and water frequently during flu season; avoiding touching the eyes or face before washing the hands; disposing promptly of soiled tissues; and not sharing personal items with other family members during a flu outbreak. The use of alcohol-based sanitizers on work surfaces and kitchen counters may be helpful in some households.

The most effective means of preventing flu is vaccination. There were two forms of vaccine as of 2008, an injectable vaccine given in the upper arm, and a liquid called FluMist that can be squirted or sprayed into the nose. The flu shot takes about two weeks to produce immunity. It must be given every year in order to protect the person against the newest mutation of the flu virus. The CDC publishes guidelines each year of people who should be vaccinated against flu; these include the elderly, people with HIV infection, those with diabetes and other chronic health conditions, health care workers, young children, and women who are more than fourteen weeks pregnant during flu season. People in North America who should consider vaccination should plan to be vaccinated in September or early October, before the start of the flu season.

# WORDS TO KNOW

**Pandemic:** A disease epidemic that spreads over a wide geographical area and affects a large proportion of the population.

**Reye syndrome:** A rare but potentially fatal illness that is linked to the use of aspirin in children.

# The Future

The high infectiousness of flu viruses, their ability to move back and forth between humans and other animals, and their rapid rate of change mean that it is unlikely that humans will eradicate them any time soon. One promising development, however, is research on a universal vaccine against influenza A viruses. Such a vaccine would eliminate the need to reformulate flu vaccines each year, which is a slow and inefficient process. One such universal vaccine had completed Phase I clinical trials as of early 2008.

SEE ALSO Common cold; Pneumonia; Reye syndrome

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## m

## Iron-Deficiency Anemia

#### Definition

Anemia is a condition in which the blood does not have enough hemoglobin in its red cells. It is a sign of a disease process of some kind rather than a disease by itself. Anemia is the most common blood disorder in the general population. There are many different types of anemias. Some result from blood loss, some from overly rapid destruction of red blood cells (RBCs), and some from the body's failure to produce enough RBCs.

Iron-deficiency anemia is the most common form of anemia. It occurs when people do not get enough iron in their diets, when their bodies do not absorb enough iron from the diet, or when blood loss occurs. Iron is needed for the production of hemoglobin, a substance found in red blood cells that carries oxygen from the lungs to other parts of the body. When there is not enough hemoglobin in the blood, the body's tissues and organs become oxygen-starved, leading to tiredness, loss of a healthy color in the skin, headaches, and other symptoms.

#### Description

Iron-deficiency anemia varies in severity from person to person. Some people may not have any symptoms—particularly if the loss of iron happens gradually over a long period of time—and may be diagnosed in the course of a routine blood test. Others may tire easily, feel weak, lose weight, or have frequent headaches. Young children and pregnant women can develop serious health problems as a result of iron-deficiency anemia. Children with anemia may develop heart murmurs or experience delays in growth and development. Pregnant women with iron-deficiency anemia have a higher risk of giving birth prematurely or having babies with low birth weight.

Another health problem that can arise as a result of iron-deficiency anemia is a rapid or irregular heartbeat. When the blood is not carrying enough oxygen to meet the needs of the body, the heart will speed up in order to pump more blood to the tissues. Over time, the increased workload on the heart can lead to enlargement of the heart, chest pain, and even heart failure.

#### Also Known As

Iron-poor blood, sideropenic anemia

#### Cause

Not enough hemoglobin in the blood

#### Symptoms

Fatigue, pale complexion, fast heartbeat, headaches, cold hands and feet

#### Duration

Months to years unless corrected

Red blood cells from a patient with iron deficiency anemia. The cells should all be bright red. JOAQUIN CARRILLO FARGA / PHOTO RESEARCHERS, INC.



#### **Demographics**

The exact number of people in the United States with iron-deficiency anemia is not known. One estimate is 3.4 million adults and children; however, many researchers think that this figure is much too low. Irondeficiency anemia in developed countries is most common in women of childbearing age because women lose more iron than men through menstruation and pregnancy. Whereas a healthy male loses about 1 milligram of iron each day, mostly through the digestive tract, women lose between 4 and 100 milligrams of iron with each menstrual period and 500 milligrams of iron with each pregnancy. Most women need to absorb twice as much iron from their diet as men.

In developing countries, iron-deficiency anemia is six to eight times more common than in North America and Europe. One reason for the difference is that people in these countries eat less meat. It is easier for the body to absorb the iron it needs from meat than from vegetables. A second reason is that intestinal parasites—particularly hookworms—are relatively common in developing countries. Intestinal parasites can lead to iron-deficiency anemia because they cause the infected person to lose blood through the digestive tract.

## When the Doctor Recommends Iron Pills

Iron supplements can cause heartburn, constipation, or stomach cramps. To lower the risk of these side effects, people who are taking iron tablets or capsules as treatment for iron-deficiency anemia may want to try the following tips:

- Take the pills or capsules with food.
- Do not take the iron supplement at bedtime if it causes stomach upset.
- Add foods rich in fiber to the diet or take a stool softener if constipation is a problem.
- Start with a low dose of the iron supplement—perhaps just one pill or capsule per day—and work up gradually to the full dose recommended by the doctor.
- If one type of iron formula causes problems, talk to the doctor about a different formula. Some people find that iron in capsule form is easier on their digestion than pills.

Risk factors for iron-deficiency anemia include:

- Sex. Twenty percent of women of childbearing age have iron-deficiency anemia compared to 2 percent of men.
- Age. Infants or children who are not getting enough iron in their diet are at risk, particularly those who are given cow's milk rather than being breastfed and older children going through rapid growth spurts. Elderly people are also at increased risk of anemia because aging reduces the ability to taste food, leading to loss of appetite and possible malnutrition.
- Premature or low-birth-weight babies.
- Eating a strict vegetarian diet.
- Undiagnosed peptic ulcers, colon cancer, or other diseases that can cause loss of blood through the digestive tract.
- Kidney disorders that must be treated with dialysis. The process of dialysis shortens the life span of red blood cells.

#### **Causes and Symptoms**

Iron-deficiency anemia can be caused by a lack of iron in the diet, by the body's inability to absorb iron, or by the loss of blood through the digestive tract, menstruation, or pregnancy. People's diets may not contain enough iron because they are malnourished in general or because they do not eat meat, fish, or dairy products. Diseases that affect the small intestine, such as celiac disease or Crohn's disease, are the most common cause of failure to absorb iron, because the small intestine is the part of the digestive system where iron is taken into the bloodstream from partially digested food. Blood loss through the digestive tract may have several causes, ranging from colorectal cancer or peptic ulcers to intestinal parasites or long-term use of aspirin.

People with mild anemia may not have any symptoms. The most common symptoms that appear in persons with moderate

iron-deficiency anemia are tiring easily, pale skin, weakness, shortness of breath, headache, lightheadedness, and cold hands or feet. Less common symptoms include:

- · Loss of appetite and unintended weight loss
- Sore tongue
- Brittle fingernails
- Unusual cravings for ice, starch, dirt, or other substances that are not food (a condition called pica)
- Bloody or tarry-looking stools

#### Diagnosis

Iron-deficiency anemia is often diagnosed as the result of a routine blood test during blood donation or a yearly medical checkup. The blood test may reveal either that there is a low level of hemoglobin in the person's blood cells or that the blood has a low hematocrit (a measurement of the ratio of red blood cells to liquid serum in the person's blood). Normal hemoglobin levels for the general population are between 12 and 16 grams per deciliter (g/dL) of blood. A normal hematocrit level is between 36 and 46 percent for women and 46 to 56 percent for men. A patient who has a hemoglobin level below 12 g/dL or a hematocrit below 36 in women or 46 in men will be diagnosed as having iron-deficiency anemia.

Other measurements that may be taken from a blood sample are the amount of iron itself in the blood serum; the amount of ferritin, a protein that helps the body store iron; and the level of transferrin, a protein that carries iron in the blood. A low level of ferritin and a high level of transferrin point to a diagnosis of iron-deficiency anemia.

A colonoscopy or an ultrasound study may be ordered if the doctor suspects that the patient's anemia is caused by blood loss through the digestive system.

#### Treatment

The usual treatment for iron-deficiency anemia caused by an iron-poor diet is a combination of adding iron-rich foods to the patient's diet plus dietary supplements containing iron. Red meat, especially beef and liver, is the best source of iron in the diet. Other iron-rich foods include chicken, turkey, pork, fish, and shellfish; eggs; pasta or cereals that are fortified with iron; beans and nuts; dried fruits; and green leafy vegetables like spinach. The doctor may also recommend adding citrus fruits and other fruits and vegetables rich in vitamin C (mangos, apricots, strawberries, cantaloupe, watermelon, broccoli, tomatoes, peppers, cabbage, and potatoes) to the daily menu. Vitamin C helps the body absorb the iron that is present in meat and other foods.

Dietary supplements may include multivitamins containing iron and vitamin C, or iron compounds in pill or capsule form to be taken with meals. Because iron supplements can cause constipation or heartburn, patients may need to make some adjustments at mealtimes (see sidebar).

Patients whose anemia is caused by intestinal parasites will be given medications to kill the parasites. Those with colorectal cancer or other disorders of the small intestine are usually treated with surgery. Peptic ulcers are treated with a combination of antibiotics and Pepto-Bismol or drugs that block the production of stomach acid.

Patients with severe anemia may require hospital treatment, including blood transfusions and iron injections.

#### Prognosis

Iron-deficiency anemia is easily treated and has an excellent prognosis when the patient's only problem is lack of iron in the diet. Most people's hematocrit will return to normal after two months of iron therapy. Iron supplements should be continued for another six to twelve months, however, to build up the body's reserves of iron in the bone marrow. In general, older adults take longer to regain normal hemoglobin and hematocrit levels than younger people. The patients with the poorest prognosis are those whose anemia is caused by colorectal cancer.

#### Prevention

Iron-deficiency anemia can usually be prevented simply by eating a wellbalanced diet that includes iron-rich foods and foods containing vitamin C. Other preventive measures include:

- Taking iron supplements if following a vegetarian diet.
- Avoiding extreme weight-reduction diets or food fads.
- Breastfeeding a baby for the first year of life rather than giving cow's milk.
- Women who are pregnant or menstruate heavily should check with their doctor about the possible need for iron supplements as well as eating an iron-rich diet.

#### WORDS TO KNOW

**Anemia:** A condition in which a person's blood does not have enough volume, enough red blood cells, or enough hemoglobin in the cells to keep body tissues supplied with oxygen.

**Hematocrit:** The proportion of blood volume occupied by red blood cells.

- **Hemoglobin:** An iron-containing protein in red blood cells that carries oxygen from the lungs to the rest of the body.
- **Pica:** An abnormal craving for substances that are not normally considered food, like soil, chalk, paper, or ice cubes.
- Older adults should also have periodic blood tests, particularly if they take high doses of aspirin or other over-the-counter pain relievers for arthritis, or if they are losing their appetite for food.

#### **The Future**

Iron-deficiency anemia is likely to continue to be a common health problem in all age groups in the United States because it has so many different potential causes, ranging from poor diet to stomach and intestinal disorders. Researchers are presently trying to develop simpler tests for diagnosing the causes of iron-deficiency anemia. They are studying the safety of a newer iron-containing formula that would be easier to digest than current pills and capsules.

**SEE ALSO** Celiac disease; Colorectal cancer; Crohn disease; Heart failure; Lead poisoning; Prematurity; Restless legs syndrome; Sickle cell anemia; Thalassemia; Ulcers

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## u Irritable Bowel Syndrome

#### Definition

Irritable bowel syndrome or IBS is a chronic (long-term) disorder of the digestive tract characterized by changes in bowel habits—constipation, diarrhea, or a combination of both—along with abdominal cramps and bloating or gassiness. It is not contagious, inherited, or a forerunner of cancer.

#### Description

IBS is a functional disorder of the digestive tract, which means that it affects the workings of the digestive system rather than its structure. IBS is a common disorder around the world. People with IBS experience cramping or bloating in the abdomen after a meal followed by an urgent need to defecate. The patient typically notices that the stools are more frequent, looser, and may contain mucus. There are four basic patterns: the patient has mostly diarrhea; the patient is mostly constipated; the constipation alternates with diarrhea; or the patient has a mixture of the two conditions. About 75 percent of patients change from one subtype to another within a year, however.

#### Also Known As

IBS, mucous colitis, nervous indigestion, spastic colon

#### Cause

Unknown; may be triggered by infection or emotional stress

#### Symptoms

Abdominal cramps, constipation, diarrhea, gassiness

Duration Years



Image of irritable bowel syndrome, in which the intestine in the center is suffering from cramping. DAVID GIFFORD / PHOTO RESEARCHERS, INC.

IBS is a chronic rather than an acute disorder with rapid onset. It does not usually get worse over time and is not accompanied by weight loss, loss of appetite, fever, or blood in the stools. Patients with these symptoms usually have other digestive disorders.

Patients with IBS are likely to have certain other disorders at the same time. These include fibromyalgia, lactose intolerance, food allergies, migraines or other recurrent headaches, depression, and backache.

#### **Demographics**

IBS is common in the general American population, affecting as many as a fifth of the adult population. Only 10 to 20 percent of people with symptoms of IBS consult a doctor, however. Most of those seeking treatment are adults but many report that their symptoms began in childhood. Half of all patients diagnosed with IBS state that their symptoms started before age thirty-five. Patients who are over forty when their symptoms begin are less likely to have IBS and more likely to have another digestive problem.

The gender ratio of IBS varies from country to country. In the United States and Europe, women are two to three times more likely than men to have the disorder, but in India, 70 percent of IBS patients are men. The reasons for this difference are not yet known. Some doctors in the United States think that women are more likely than men to seek help for their symptoms rather than being more likely to develop it. IBS is thought to be equally common in all racial and ethnic groups in Europe and the United States, although some researchers state that the disorder is less common among Asian Americans.

#### **Causes and Symptoms**

The cause of IBS is not known. There are, however, several theories about the possible causes of the disorder. These include:

- Infections. Some researchers think that IBS may be caused by a bacterial infection. Evidence for this includes signs of inflammation in the small bowel of some patients with IBS as well as the fact that some patients diagnosed with IBS did not have symptoms until they had a gastrointestinal infection.
- Emotional trauma. Some studies indicate that a significant number of women diagnosed with IBS are survivors of physical or sexual abuse.
- Abnormal patterns of contraction of the muscles in the walls of the intestines. During the process of digestion, the muscular walls of the intestines push food along the digestive tract by rhythmic contractions. In some patients with IBS, the contractions are too close together or too far apart, leading to the cramping sensations and diarrhea or constipation of IBS.
- Unusual sensitivity of the nerve endings in the intestines. Some researchers think that the intestinal tissues in patients with IBS are more sensitive to stretching than those in most people, or that there are more intense connections between the central nervous system and the intestines in patients with IBS.

In addition to cramping, bloating, and changes in bowel habits, patients with IBS may have the following symptoms:

- Feeling a need to defecate even when there is nothing in the bowel or rectum.
- Feeling that the bowel has not been completely emptied after a movement.
- Finding that cramps and gassiness are relieved by a bowel movement.
- Visible swelling or bloating of the abdomen.
- Finding mucus in the stools.

#### Diagnosis

IBS is a diagnosis of exclusion, which means that the doctor must rule out other possible causes of the symptoms. There is no single laboratory test or imaging study that can confirm a diagnosis of IBS. There are several sets of diagnostic criteria drawn up by various professional groups that the doctor can use. Most of these criteria state that the patients must have had abdominal pain or bloating for at least twelve weeks in the past year; that the pain is relieved by a bowel movement; and that the patient has noticed changes in the shape, frequency, or appearance of the stools.

If the patient is over fifty, the doctor may order tests to rule out the possibility of colon cancer. Other tests may be ordered if the patient has fever, weight loss, blood in the stools, or persistent pain, as these symptoms are not characteristic of IBS.

#### Treatment

There is no cure for IBS. Patients may be treated with a combination of medications, dietary adjustments (described more fully in the section on prevention), and psychotherapy. In some cases the doctor may ask the patient to keep a food diary to see whether certain foods make the symptoms worse. Patient education is a very important part of treatment for this disorder, as there are steps that patients can take to manage their symptoms and reduce the frequency of flare-ups.

The specific medications prescribed depend on the patient's most bothersome symptoms. They may include:

- Fiber supplements like Metamucil or Citrucel. These are taken with fluids to relieve constipation.
- Antispasmodic drugs. These are given to slow down the contractions of the intestines. They include drugs like Bentyn and Levsin.
- Antidepressants. Drugs like Paxil and Prozac are reported to help patients with severe constipation. They are also given to IBS patients with coexisting depression.
- Antidiarrheal medications. Imodium and Lomotil are the drugs most often recommended for treating severe diarrhea.
- There are also some newer drugs that are available only for patients who do not respond to other treatments. Lotronex and Tegaserod are drugs approved only for temporary use in women with severe IBS symptoms. Both drugs have potentially serious side effects and

have not been approved by the Food and Drug Administration (FDA) for treating men.

There are no surgical treatments for IBS. Some alternative therapies that some patients find helpful include acupuncture and herbal remedies, particularly peppermint tea or capsules. Peppermint is a natural antispasmodic that relaxes the intestinal muscles.

#### Prognosis

IBS is a bothersome condition but it is not life-threatening and will not cause or lead to cancer. Patients with IBS have the same life expectancy as others of their age or sex in the general population.

#### Prevention

People with IBS cannot completely prevent occasional episodes of diarrhea or constipation with any medications presently available, but they can minimize the severity of their symptoms in a number of ways:

- Psychotherapy or counseling. Some people find their symptoms are helped by learning to avoid overreacting to normal life stressors.
- Getting regular exercise. Exercise helps to maintain bowel function as well as lower stress levels.
- Avoiding foods that produce cramping or gas. These include coffee, spicy foods, beans, onions, broccoli, and cabbage.
- Practicing eating slowly and avoiding overeating.
- Quitting smoking and reducing or eliminating alcoholic beverages.
- Practicing yoga, meditation, relaxation techniques, or deep breathing.
- Cutting back on cola drinks and other carbonated beverages.
- Adding wheat bran or other foods high in fiber to the diet.
- Hypnosis. Some patients with IBS report significant symptom relief when they are taught self-hypnosis aimed at relaxing the muscles of the abdomen.

#### The Future

IBS is likely to continue to be a common problem among American adults. Current research includes clinical trials of several new drugs for IBS; investigations of alternative therapies, including traditional Chinese

#### WORDS TO KNOW

**Antispasmodic:** A type of drug given to relieve the cramping of the intestines or other muscles.

**Lactose intolerance:** An inability to digest lactose, the form of sugar found in milk and milk products.

medicine, St. John's wort (a herbal remedy), and massage therapy; and imaging studies to see whether the brains of patients with IBS are different from those of people without the disorder.

**SEE ALSO** Celiac disease; Crohn disease; Fibromyalgia; Lactose intolerance; Ulcerative colitis

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# K



Genetic

Infection

Injury

Multiple

Other

Unknown

Also Known As XXY syndrome; 47, XXY syndrome

#### Cause

An extra X chromosome in a male's genetic makeup

#### Symptoms

Small genitals, sparse body and facial hair, enlarged breasts, unusual height, infertility

#### Duration

Lifelong



## Klinefelter Syndrome

#### Definition

Klinefelter syndrome is a condition caused by one or more extra X chromosomes in males. It is the second most common health condition caused by an extra sex chromosome. Klinefelter syndrome is named for Dr. Harry Fitch Klinefelter (1912–1990), an endocrinologist at Massachusetts General Hospital in Boston, who first described it in 1942. The genetic cause of the syndrome was not discovered until 1959.

#### Description

Many men with an XXY chromosome arrangement tend to be taller than their father or brothers, to have a rounded body type with a tendency to be overweight, to develop enlarged breasts resembling those of a woman, to have a smaller than average penis and testicles, and to lack facial or body hair. On the other hand, many other men who are XXY do not develop these features; some live out their lives without ever knowing that they have an extra X chromosome. For this reason, many doctors no longer use the term "Klinefelter syndrome" but prefer to describe males with the extra chromosome simply as "XXY males."

#### Demographics

Klinefelter syndrome affects only males. It is thought to occur in one in every 500–1,000 boy babies. About 3,000 affected males are born each



Chromosomes of a person with Klinefelter syndrome. There is an extra X chromosome, resulting in 47 chromosomes, rather than the normal 46. CNRI / PHOTO RESEARCHERS, INC.

year in the United States. The rate of Klinefelter syndrome is five to twenty times higher in boys with mental retardation than in the general population of newborns. It appears to be equally common in all races and ethnic groups.

#### **Causes and Symptoms**

Klinefelter syndrome is caused by a type of genetic error called nondisjunction. Ordinarily, the process of cell division that leads to the formation of germ cells (sperm and eggs) results in a cell with only half the number of chromosomes twenty-three, instead of forty-six. In nondisjunction, the division of sex chromosomes does not occur, resulting in either an egg with two X chromosomes or a sperm with both an X and a Y chromosome. If a normal sperm carrying a Y sex chromosome fertilizes an egg with two Xs, or a sperm carrying both an X and a Y chromosome fertilizes a normal egg, an XXY male will be con-

ceived. According to researchers, about half the time the extra chromosome comes from the baby's father and the other half of the time from the mother. A mother who is thirty or older has a slightly increased risk of having an XXY son.

Klinefelter syndrome is characterized by a range of psychosocial as well as physical problems. Not all XXY males will have all these difficulties, and many have them only in mild form.

- Smaller than normal testes and penis, with lower than normal levels of male sexual hormones.
- Thin or absent body and facial hair.
- High-pitched voice.
- Rounded body shape, enlarged breasts, rapid weight gain in adolescence.
- Developmental and learning disabilities. These include delayed speech, difficulty paying attention, mild difficulties with short-term memory, and problems with learning to read.
- Depression and emotional distress caused by low-self-esteem.

- Taller than average height after puberty with disproportionately long arms and legs.
- Infertility (inability to father children).

XXY males are at increased risk of certain autoimmune disorders, including rheumatoid arthritis and lupus. They are also at increased risk of breast cancer and osteoporosis.

#### Diagnosis

Diagnosis of Klinefelter syndrome depends in part on the severity of the patient's symptoms. As has been noted, many XXY males are only mildly affected by their extra X chromosome and may never be diagnosed. Although babies can be diagnosed with the syndrome before birth, the two most common symptoms that bring XXY males to the doctor's office for testing are enlarged breasts and infertility, both of which become matters of concern after puberty. Some XXY boys are diagnosed during the elementary school years because they have speech problems and other learning difficulties.

The diagnosis is made by taking a karyotype, or chromosome test. To perform a karyotype, the doctor takes a small sample of the patient's blood. The white blood cells are isolated and cultured in a special solution and examined under a microscope to see what the chromosomes look like.

The doctor may also test a sample of blood for hormone levels. XXY males have low levels of testosterone, a male sex hormone, in their blood serum.

### One Man's Story

An XXY male who was born in 1961 and grew up in the 1970s tells about the difference that having a karyotype (chromosome test) made in his life. He was diagnosed with Klinefelter syndrome in junior college. Before his diagnosis, he had problems in school related to low energy levels as well as gaining large amounts of weight: "The major problem faced by me, and by every other Klinefelter's Syndrome victim, is that maturity and learning go from normal to very, very slow after puberty kicks into action. It takes energy to learn, and I didn't have any.... I could not keep up physically, nor could I keep up intellectually. In some things, like computers, I worked well, for I could sit in one place and learn. If I had to move around, go visit the library, do research, exercise, or make any effort at all, I was muted by a weakness that was always labeled as fat and lazy."

After testosterone therapy, the writer had much more energy, joined his parents in their business for several years, was able to return to college eventually and complete his degree in 2003. He married and was able to lose weight after obesity surgery. He summarizes the benefits of his diagnosis and treatment by reaching out to other XXY males: "I keep meeting people with Klinefelter's who are not taking testosterone and remaining weak and immature in many ways. Don't avoid testosterone. It matured more than my physical form. It matured my mind."

#### Treatment

Treatment of Klinefelter syndrome may involve an educational evaluation as well as medical treatment:

• Hormone injections. Injections of androgens (male sex hormones) are given to XXY males, preferably beginning at puberty, in order

to help them gain muscle strength, develop facial hair and a deeper voice, enlarge the testes, raise overall energy levels, and protect against osteoporosis. In many cases hormone treatment improves the boy's mood and self-esteem as well.

- Surgery. XXY males with noticeably enlarged breasts may have surgery to remove the extra breast tissue. Surgical treatment reduces the man's risk of breast cancer as well as removing a cause of social embarrassment.
- Speech therapy and language therapy. Most doctors recommend that XXY boys have a complete educational evaluation, preferably in elementary school, so that their learning difficulties (if any) can be treated before they develop behavioral problems or become depressed.
- Physical therapy. Some XXY boys benefit from exercises that help them improve their muscle strength and coordination.

Most doctors consider mid-to-late adolescence the best time to tell an XXY boy about his condition. At that age most are able to understand the cause of the syndrome and its implications, and to decide whether they want to share the information with anyone else.

#### Prognosis

Most XXY males can live productive lives with normal life expectancy; many complete college and graduate school. In 1996 a technique was developed for extracting sperm from the male testicle and injecting it into a female egg; since that date, at least sixty children around the world have been conceived and born using sperm from men with Klinefelter syndrome. Men who are able to father children this way do not have any greater risk of producing an XXY son than men in the general population.

#### Prevention

There is no known way to prevent Klinefelter syndrome because it is caused by a random genetic error.

#### The Future

It is unlikely that Klinefelter syndrome will become more common in the general population in the future because nondisjunction is a random

#### WORDS TO KNOW

- **Androgen:** The generic term for the group of male sex hormones produced by the body.
- **Germ cell:** A cell involved in reproduction. In humans the germ cells are the sperm (male) and egg (female). Unlike other cells in the body, germ cells contain only half the standard number of chromosomes.
- **Karyotype:** A photomicrograph of the chromosomes in a single human cell. Making a karyotype is one way to test for genetic disorders.
- **Nondisjunction:** A genetic error in which one or more pairs of chromosomes fail to separate during the formation of germ cells, with the result that both chromosomes are carried to one daughter cell and none to the other. If an egg or sperm with a paired set of chromosomes is involved in the conception of a child, the child will have three chromosomes in its genetic makeup, two from one parent and one from the other.

Testosterone: The principal male sex hormone.

genetic error. What is likely, however, is that earlier diagnosis and better understanding of the syndrome will help XXY males do well in school and the adult workplace and lower their risk of depression and other setbacks. The knowledge that most XXY males benefit from hormonal treatment, surgery, and various supportive educational measures and can have normal lives is certainly encouraging.

SEE ALSO Breast cancer; Lupus; Osteoporosis; Rheumatoid arthritis

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Genetic

Infection

Multiple

Injury

Other

Unknown

Also Known As Lactase deficiency

#### Cause

Underproduction of lactase, a digestive enzyme

#### Symptoms

Intestinal gas, bloating, nausea, and diarrhea

#### Duration

May last a few weeks or be lifelong

### **m** Lactose Intolerance

#### Definition

Lactose intolerance occurs when a person cannot digest lactose, a sugar found in milk and other dairy products. Lactose intolerance develops when lactase, an enzyme that is needed to break down milk sugar into simpler sugars, is less available or absent.

#### Description

Lactose intolerance is a very common chronic digestive disorder in which a person's intestinal tract lacks the ability to make lactase, an enzyme that breaks down lactose, or milk sugar, into two simpler sugars that the body can use. A person can have a lactase deficiency without having the symptoms of lactose intolerance.

Lactose intolerance is not the same as being allergic to cow's milk. An allergy to cow's milk concerns a person's immune system, whereas lactose intolerance has to do with the process of digestion.

Lactose intolerance may be caused by any of three different factors. One is normal aging. As people get older, their small intestine produces lower amounts of lactase. After the lactase production drops below a certain point, the person may experience the symptoms of lactose intolerance.

A second cause of lactose intolerance is diseases of the intestines or surgical procedures in which part of the small intestine is removed. These

#### Foods to Watch

Foods that are naturally high in lactose (in addition to milk) include butter and sometimes margarine; buttermilk; cottage cheese, cream cheese, and ricotta cheese; half and half, light cream, and whipping cream; ice milk, sherbet, ice cream, evaporated milk, and dry powdered milk; milk chocolate; whey; and yogurt.

Foods that contain hidden lactose include hot dogs, cold cuts, bologna, sausages, pancakes, creamy salad dressings, creamed soups, breaded meats, commercial pie crust and pie fillings, caramels, fudge and other chocolate candies, prepared cakes and sweet rolls, powdered coffee creamers, imitation dairy products, party dips, cream-based cordials, certain breads, sauces and gravies, frosting, certain prepared or processed foods, some prescription medications, and some over-the-counter medications. disorders or operations may affect the part of the small intestine that secretes lactase. The third cause of lactose intolerance is genetic. A few people inherit lactose intolerance from both parents and are affected from birth.

The symptoms of lactose intolerance usually begin within half an hour to two hours after drinking milk or eating a meal high in dairy products. The person typically experiences diarrhea, which is the most common symptom of lactose intolerance, along with a gassy, bloated feeling, abdominal cramps, and possibly nausea. The severity of the symptoms is not necessarily related to the amount of milk or dairy products that were consumed but rather to the person's age, ethnicity, and the speed of his or her digestive processes.

#### Demographics

In most cases, lactose intolerance is part of the normal human developmental process. Most mammals stop producing lactase after they are

weaned because they are eating solid food instead of drinking milk from the mother. Humans begin to slow down the production of lactase some time around age three to five years; thus most human adults are at some risk of developing lactose intolerance.

It is noteworthy, however, that the levels of lactose intolerance vary quite widely among different ethnic groups. In some groups, almost 100 percent of the adult population may be lactose intolerant. In the United States and Canada, lactose intolerance is estimated to affect between 20 and 60 percent of the adult population. In terms of specific ethnic groups, people of Dutch, Swedish, German, or other northern European descent have low rates of lactose intolerance (about 5 percent); persons of southern European ancestry have rates between 18 and 25 percent; African Americans have a rate around 45 percent; persons from Japan or southeastern Asia have rates above 95 percent; and Native Americans are almost 100 percent lactose intolerant. One theory that has been proposed to explain these differences is the long-standing differences among human societies in milk consumption after childhood. In Asia and Africa, children were rarely given milk after being weaned; in these societies, lactase production generally falls by 90 percent by the time the child is four years old. In societies in which milk consumption continues into adult life, however, a mutation on chromosome 2 that bypasses the normal shutdown of lactase production became widespread in the population. Thus members of these groups can continue to consume milk and dairy products throughout their adult lives. Some researchers have traced the mutation back as far as 4500 BCE in both Sweden and the Middle East.

#### **Causes and Symptoms**

Lactose intolerance results from a drop in or disruption of the production of lactase in the small intestine. Lactase is produced by specialized cells in the membrane that lines the villi, which are small finger-like projections on the walls of the small intestine. The production of the enzyme may drop at a certain age or because a disease or radiation treatment for cancer has damaged the villi of the small intestine.

The symptoms of lactose intolerance are diarrhea, bloating, nausea, and a gassy feeling within thirty minutes to two hours following a meal high in dairy products. They do *not* include fever, bleeding from the digestive tract, or weight loss in adults. People who have these symptoms should be checked by their doctors for other disorders of the intestines.

#### Diagnosis

Diagnosis of lactose intolerance is based on a patient's history, particularly a detailed history of the patient's consumption of dairy products. Many people underestimate the amount of milk or products containing lactose that they consume; they may not think of yogurt or ice cream, for example, as milk products. After getting a complete picture of the patient's diet, the doctor will usually suggest cutting out dairy products for a week or so in order to see whether the symptoms improve. If they do, further testing may be unnecessary.

There are three tests that can be used, one of which is generally given only to infants and small children. It is a test that measures the acidity of the child's stool sample. Undigested lactose ferments inside the intestine and forms an acid that can be measured in the stool sample. The most common diagnostic test used in adults is the hydrogen breath test. The patient is asked to drink a liquid containing a high level of lactose. The doctor then measures the amount of hydrogen in the breath at certain intervals. Undigested lactose reaches the colon and ferments, causing hydrogen and other gases to be released, absorbed by the intestines, and eventually exhaled. Large amounts of exhaled hydrogen indicate that the patient's body is not digesting lactose completely and that the patient is probably lactose intolerant.

The third type of diagnostic test involves taking a small sample of tissue from the lining of the small intestine and measuring the amount of lactase present in the tissue sample. This type of test requires a specialized laboratory to evaluate the results, however, and is rarely used outside clinical research.

#### Treatment

There are several treatment options for lactose intolerance:

- Completely eliminating milk and dairy products from the diet. This change in diet usually requires careful reading of labels on other foods because many processed foods contain milk or milk solids (see sidebar). In addition, some drug manufacturers use lactose as a binding substance to carry the active ingredient in the medication. The patient may need to check with the doctor or pharmacist about any prescription medications they may be using to see if the drugs were formulated with lactose.
- Eliminating dairy products from the diet for a time and then gradually reintroducing small amounts of them. Some people can tolerate small amounts of yogurt or milk after avoiding them completely for a few weeks.
- Using specially manufactured lactose-free milk products or soy products and other plant-based substitutes for milk.
- Taking dietary supplements that contain lactase. Products like Lactaid, DairyEase, and Lactogest can be purchased without a prescription.

People who are concerned about the risk of osteoporosis (brittle bones) can take calcium supplements to keep their bones strong rather than getting their calcium from milk. Patients should ask their doctors how much calcium they should be getting from other sources. Most adults should not take more than 1,200–1,500 milligrams of calcium per day.

#### WORDS TO KNOW

Chronic: Recurrent or long-term

Congenital: Present from birth.

**Lactase:** An enzyme that breaks down lactose into simpler sugars during the process of digestion.

Lactose: A complex sugar found in milk and other dairy products. It is sometimes called milk sugar.

Villi (singular, villus): Small finger-like projections along the walls of the small intestine that increase the surface area of the intestinal wall.

#### Prognosis

Most people recover completely by removing milk products from the diet or by substituting reduced-lactose or lactose-free dairy products for those that contain lactose.

#### Prevention

There is no way to prevent congenital or adult-onset lactose intolerance.

#### The Future

Lactose intolerance is not a life-threatening condition. Most people can manage quite well by using milk substitutes, watching the amount of milk and other dairy products that they consume, or by taking overthe-counter lactase supplements. There are also a number of cookbooks with lactose-free recipes or recipes that use milk substitutes.

**SEE ALSO** Celiac disease; Crohn disease; Irritable bowel syndrome; Osteoporosis

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## m

### Laryngitis

#### Definition

Laryngitis is defined as inflammation or irritation of the larynx, which is the voice box that lies at the base of the throat just above the windpipe. It is a condition or symptom rather than a distinctive disease. Acute laryngitis is defined as lasting three weeks or less; laryngitis that lasts longer than three weeks is called chronic laryngitis.

#### Description

Laryngitis occurs when the vocal folds (or vocal cords) swell as a result of infection or another cause of inflammation. The vocal folds are two bands of tissue that stretch across the larynx. Under normal circumstances, when a person wants to speak, the vocal folds tighten. Air from the lungs is forced through the smaller space between the bands of muscle, causing them to vibrate. Lengthening, shortening, tightening, and loosening of the cords allows a person to control the pitch of the voice. When the vocal folds swell up due to irritation or inflammation, they cannot vibrate easily, which causes the voice to sound hoarse, raspy, or faint.

#### Also Known As

Losing one's voice, dysphonia

#### Cause

Overuse of the voice, infection, or throat irritation

#### Symptoms

Hoarseness, difficulty speaking, sore throat, dry cough

#### Duration

Seven to ten days (acute); over three weeks (chronic)

#### Demographics

Laryngitis is so common in the general population that no exact statistics are kept. Most people treat acute laryngitis at home without visiting a doctor, particularly if the voice problem seems to be a side effect of a cold or the flu. According to one study, acute laryngitis is most common in adults between the ages of eighteen and forty. It appears to affect both sexes and all races equally. Chronic laryngitis is more common in adults over fifty and in people whose occupations expose them to irritating chemicals.

People who smoke, people with asthma, firefighters, and singers or public speakers are at greater risk of laryngitis than the general population.

#### Home Care for Laryngitis

Acute laryngitis caused by a cold or the flu can usually be treated at home:

- Keep the air in the house moist by using a humidifier.
- Moisten the tissues of the throat by breathing in the steam from a hot shower or holding the heat over a bowl of hot steaming water.
- Drink plenty of fluids.
- Use throat lozenges, a salt-water gargle, or chewing gum to help keep the throat moist.
- Give the voice as much complete rest as possible.
- Avoid whispering; whispering is harder on the vocal folds than normal speech.

#### **Causes and Symptoms**

The most common single cause of acute laryngitis is an upper respiratory infection caused by a virus, most often a cold or influenza virus. Other viruses, such as those that cause chickenpox, mumps, or measles, can also cause laryngitis.

Other causes of laryngitis include:

- Infections caused by bacteria or fungi
- Irritation of the throat caused by smoking
- Drying of the tissues lining the throat caused by asthma inhalers, overuse of decongestants, or antihistamines
- Exposure to dust, chemicals, smoke, fumes, or other irritating substances in a person's workplace
- High levels of alcohol consumption
- Air pollution
- Gastroesophageal reflux disease (GERD; a disorder in which acid from the stomach flows backward up the esophagus and into the throat, irritating the throat tissues.)
- Repeated episodes of sinus infection
- Throat cancer

In addition to a hoarse or faint voice, people with laryngitis may complain of soreness in the throat, a tickling sensation, a dry cough, difficulty breathing, or discomfort when swallowing food.

#### Diagnosis

Acute laryngitis is usually diagnosed by taking the patient's history particularly recent exposure to colds or flu—and an examination of the throat and neck. The doctor will usually feel the outside of the neck for signs of swollen lymph glands and will look down the patient's throat using a mirror or with a device called a laryngoscope. If the patient has acute laryngitis, the vocal folds will look red, swollen, and covered with fluid secretions. Laboratory studies are not usually needed.

If the patient has chronic laryngitis, the doctor will examine him or her for signs of GERD or refer the patient to an otolaryngologist. Otolaryngologists are doctors who specialize in diagnosing disorders of the ears, nose, and throat. The patient may need some special examinations to evaluate the possibility of throat cancer.

#### Treatment

People can often treat acute laryngitis at home with some simple remedies (see sidebar). It is not usually necessary to take antibiotics for acute laryngitis, as several studies have shown that these drugs do not speed up the patient's recovery. If the laryngitis does not clear up after two weeks, however, the patient should have the throat checked again.

Chronic laryngitis caused by GERD is usually treated by drugs that lower the production of stomach acid, by changes in the patient's diet, and by raising the head of the bed during sleep. The laryngitis usually clears up once the abnormal backward flow of stomach acid into the throat stops.

Laryngitis caused by overuse of the voice is usually treated by complete vocal rest for several days. Even if the patient is a professional singer, cheerleader, or public speaker, trying to use the voice during an episode of laryngitis can make the condition worse.

#### Prognosis

Acute laryngitis caused by an infection usually clears up completely in about a week. The prognosis of chronic laryngitis depends on the cause of the condition. People with chronic laryngitis caused by GERD or by overuse of the voice will usually recover without complications if they follow the doctor's advice about diet and vocal rest. The prognosis of

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### WORDS TO KNOW

**Chronic:** Recurrent or long-lasting.

**Larynx:** The medical name for the voice box located at the base of the throat.

**Otolaryngologist:** A doctor who specializes in diagnosing and treating diseases of the ears, nose, and throat.

**Pitch:** The highness or lowness of the voice or a musical note.

**Vocal folds:** Twin folds of mucous membrane stretched across the larynx. They are also known as vocal cords.

throat cancer depends on the stage of the cancer at the time it is diagnosed. Fortunately, throat cancer is a very rare cause of laryngitis.

#### Prevention

Some measures can be taken to reduce the risk of getting laryngitis:

- Quitting smoking (or not starting in the first place) and avoiding secondhand smoke.
- Taking precautions against colds and flu; for many people, these measures include an annual flu shot.
- Avoiding the overuse of antihistamines or decongestants when treating a cold at home.
- Avoiding breathing irritating household cleansers and other chemicals.
- Avoiding overuse of the voice, including loud yelling or screaming.

#### **The Future**

Laryngitis is a common health problem that is likely to continue to be common, if only because it is so often associated with colds and other common respiratory ailments. Laryngitis related to occupational hazards is also likely to continue to be a common health problem.

**SEE ALSO** Asthma; Common cold; Gastroesophageal reflux disease; Influenza; Smoking; Sore throat

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### Lead Poisoning

#### Definition

Lead poisoning is a form of chronic (long-term) damage to the nervous system and other body organs. It is caused by inhaling dust containing lead or eating or drinking material contaminated with lead.

#### Description

Lead poisoning has been a source of human illness and premature death since people first began to use lead in pottery, plumbing, cosmetics, jewelry, metal cookware, and even medicines. The oldest known lead mine, opened about 6500 BCE, is located in present-day Turkey. Lead poisoning was first identified around 200 BCE by Nicander of Colophon, a Greek doctor. The metal was popular in the ancient world, however, because it is easily worked, it has a low melting point, and it does not rust. The most common sources of lead poisoning from ancient Rome through the Middle Ages were drinking vessels made of pewter, a metal made mostly of tin with small amounts of copper and lead added; and wine, which was often sweetened by a compound of lead called lead acetate or sugar of lead.

Lead poisoning in humans is often slow to develop because of the small quantities of the metal that can cause health problems. Lead harms the body by preventing the body from using iron, zinc, and calcium in the production of hemoglobin (a pigment found in red blood cells) and in other important body processes. In addition, lead is not easily removed from the body. After it enters the body through the lungs or the digestive tract, it travels first to the blood and other internal organs and then is stored in the bones and teeth. Lead in the blood and soft tissues is

Also Known As

Lead toxicity, plumbism

#### Cause

Inhaling or ingesting lead from paint, soil, or other objects

#### Symptoms

Muscle weakness, anemia, damage to brain and nervous system, infertility, joint pain

#### Duration

May be lifelong



*Illustration showing various sources of lead that can cause lead poisoning.* THE GALE GROUP.

gradually filtered by the kidneys over a period of sixty to seventy days but may remain in the bones and teeth for several years.

#### Demographics

In the United States, lead poisoning is most likely to affect children below the age of five years and adults in certain high-risk occupations. About 1.5 million workers are exposed to lead in the workplace. The number of children with risky blood levels of lead, or BLLs, has decreased 68 percent since 1991. As of 2002, the most recent survey year, 1.6 percent of American children between one and five years of age had BLLs above 10 micrograms of lead per deciliter (about 3.5 ounces) of blood.

Children of any race living in cities or near highways with high levels of auto exhaust fumes are at greater risk of high BLLs than children living in rural areas. African American children are at greater risk than either Hispanic or Caucasian children. Boys and girls are at equal risk.

#### **Causes and Symptoms**

The basic cause of lead poisoning is inhaling, swallowing, or otherwise being exposed to lead in paint, soil, or lead-based products in the environment. Although lead is no longer used in the manufacture of paints, gasoline, and other products, people can still be exposed to dangerous amounts of lead. In the United States, the most common sources of exposure to lead include:

- Houses built before the 1960s and painted before 1978. House paint was made with lead until 1978, but many older houses have layers of paint that were applied before lead-free paints were available. Stripping or sanding away older paint can be dangerous, too, because the removal process releases fine dust particles containing lead into the air. Children can breathe in this dust or take in lead by swallowing paint chips or dust from lead-based paint.
- Toys and furniture that were painted before 1976.
- Plumbing, pipes, and faucets.
- Storage batteries.
- Children's toys made outside the United States, including paint sets and art supplies.
- Soil contaminated by long years of car exhaust, such as soil along the sides of highways or near gas stations.
- Lead bullets, fishing sinkers, and curtain weights.
- Homemade illegal whiskey ("moonshine").
- Pewter pitchers, goblets, and dinnerware.
- Lead-based costume jewelry.
- Hobbies that involve soldering, glazing pottery, making jewelry, or making items out of stained glass.

Other sources of lead poisoning in some ethnic groups in the United States are folk remedies for various ailments. These include the practice of eating soil, certain traditional medicines imported from Southeast Asia, and a folk remedy called litargirio, sold in Hispanic grocery stores.

The symptoms of lead poisoning usually develop gradually over time rather than coming on suddenly. In many cases the symptoms are not specific to lead poisoning and may be mistaken for the symptoms of other disorders. In children, whose nervous systems are more vulnerable to lead, symptoms of lead poisoning include irritability, loss of appetite and weight, tiredness, abdominal cramps, vomiting, constipation, a pale complexion due to anemia, impulsive behavior, seizures, lowered IQ, and learning difficulties. Symptoms of lead poisoning in adults may include:

- · Pain, numbness, or tingling sensations in the arms and legs
- Difficulty conceiving in women or abnormal sperm in men
- Headache and memory problems
- Abdominal cramps and constipation
- Mood disorders and personality changes
- Muscle pains and weakness
- Hypertension (high blood pressure)
- Cataracts

#### Diagnosis

Most cases of lead poisoning are detected by screening people at risk rather than because the doctor suspects that the symptoms are caused by lead. The diagnosis of lead poisoning is based on a blood test called the blood lead level or BLL. The test can be given to measure the effects of treatment for lead poisoning as well as to screen people for exposure to harmful amounts of lead.

The blood test results are evaluated according to standards set by the American Academy of Pediatrics and the Centers for Disease Control and Prevention (CDC). The BLL is measured in micrograms of lead per deciliter of blood (mcg/dL). A deciliter is about a fifth of a pint. The definition of what is considered a dangerous level of blood lead has changed over the years. In 1997 the blood lead level of concern for children was decreased from 25 micrograms per deciliter to 10 micrograms per deciliter.

Not everyone needs to be screened for possible lead poisoning. The CDC recommends screening for adults employed in certain occupations, particularly metal working, glass working, lead plating, ore refining, auto repair, road repair, and construction. Children in the following categories should be screened for possible lead poisoning:

- Children whose families are receiving federal assistance
- Children who live in or regularly visit a house or apartment built before 1950, or before 1978 if the house has been or is undergoing remodeling
- Children who have a sibling or playmate diagnosed with lead poisoning
- Children from refugee or immigrant families

The CDC divides the results of the BLL test into six groups or classes:

- Class I: Less than 10 mcg/dL
- Class IIA: 10–14 mcg/dL
- Class IIB: 15-19 mcg/dL
- Class III: 20-44 mcg/dL
- Class IV: 45–69 mcg/dL
- Class V: 70 mcg/dL or higher (This blood lead level is considered a medical emergency.)

#### Treatment

Mild cases of lead poisoning (Classes I through III) can often be treated simply by removing the source of the lead. Children should have their BLL retested a month later to make sure their blood lead level is dropping.

People with higher BLLs are usually treated with chelation therapy. This is a type of treatment in which the person is given a drug that binds with the lead in the body so that it can be excreted in the urine. People in Class III and some in Class IV are given a drug called succimer, which is taken by mouth. People with BLLs above 50 mcg/dL are usually treated with a drug called EDTA, which must be given intravenously.

Children who have developed anemia as a result of lead poisoning may be given iron supplements.

#### Prognosis

The prognosis for recovery from lead poisoning depends on the patient's age and the level of lead in his or her body. Adults with low levels of lead often recover without problems. Those with higher levels have a greater risk of long-lasting health problems and must be monitored carefully by their doctor. Their nerves and muscles may no longer function well. More-over, other body systems may be harmed to various degrees, including the kidneys and blood vessels. People who survive toxic lead levels may suffer some permanent brain damage. However, death from lead poisoning is rare in the early 2000s because of the widespread use of chelation therapy.

Some people also develop complications after chelation therapy, as treatment with chelating drugs does not always reverse nerve damage. Depression, increased aggressiveness, impotence, and infertility have been reported in adults.

#### WORDS TO KNOW

**Chelation therapy:** Treatment of lead poisoning by administering medications that help the body excrete the lead in the urine.

**Pewter:** A metal made mostly of tin and small quantities of copper. Modern pewter is no longer made with lead.

Plumbism: The medical name for lead poisoning.

#### Prevention

The National Institute for Environmental Health Sciences (NIEHS) recommends the following measures to reduce people's exposure to lead:

- People who think they may have lead paint in their homes should get advice on safe paint removal from the Housing and Urban Development (HUD) at 800-RID-LEAD or the National Information Center at 800-LEAD-FYI.
- Parents who work in occupations where they are exposed to lead should change clothes before coming home.
- Everyone should wash their hands before meals.
- People should have the household tap water tested for lead levels. If it is high, filter it or use bottled water for drinking and cooking.
- Throw out old painted toys if it is unclear whether the paint contains lead.
- The home should be kept as dust-free as possible.
- Avoid using candies, canned goods, or folk medications produced outside the United States.

#### The Future

Lead poisoning is likely to continue to be a public health problem for some time. One reason is that lead is not biodegradable. This means that lead persists in the outside environment for a long time. There are still many older buildings that were painted with lead-based paints before 1978, and many people are still unaware of the dangers of removing old paint without safeguards. In addition, it is difficult to control the entry and sale of folk medicines containing lead in the United States.

SEE ALSO Cataracts; Depression; Hypertension

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# m

### Learning Disorders

Learning disorders is a term used to describe childhood school-related problems related to academic and functional skills, including being able to read, write, spell, speak, listen, think a problem through logically, and organize information. Learning disorders are sometimes called learning disabilities. They can be caused by a number of factors, ranging from heredity and damage to the child's brain before birth to premature birth or accidents after birth.

A learning disorder is not the same thing as mental retardation or having low intelligence. Many children diagnosed with learning disorders have average or above-average intelligence. Some doctors categorize learning disorders according to whether the child's problem is caused by:

- Problems taking in information through the senses (vision problems or hearing loss).
- Problems organizing information in the mind. This skill includes relating new information to facts previously learned and being able to put facts together to form a larger picture of the subject.
- Memory problems.
- Problems with speech or motor activities (drawing or handwriting).

Specific learning disorders include dyslexia, the most common learning disorder; writing disorder; mathematics disorder; motor (movement) skill disorder; and disorders of speaking and listening. Treatment of learning disorders is focused on identifying them and working with the child to overcome them as early as possible.

SEE ALSO Dyslexia; Fetal alcohol syndrome; Hearing loss; Prematurity

## U

### Leukemia

#### **Definition and Description**

Leukemia is the name of a group of cancers that affect white blood cells. It takes its name from the abnormally high numbers of white blood cells found in patients' blood before treatment. It is not a single disease; there are four major types of leukemia, two that are considered acute (they worsen rapidly) and two that are chronic (they progress slowly). These four types are:

• Chronic lymphocytic leukemia (CLL). A lymphocytic leukemia is one in which the cancer affects white blood cells (WBCs) called lymphocytes. The abnormal but relatively mature cells multiply, keeping normal cells from doing their job of fighting infections. Also Known As Cancer of the blood

**Cause** Unknown

#### Symptoms

Fatigue, easy bruising and bleeding, swollen lymph glands, frequent infections

**Duration** Years


Magnified image of leukemia cells in blood. © PHOTOTAKE INC. / ALAMY.

CLL is related to another type of cancer called lymphoma, which is a cancer that affects the lymphatic system. CLL is a common adult leukemia; it progresses slowly and many patients feel well for years without treatment.

• Chronic myeloid leukemia (CML). A myeloid leukemia is one that affects bone marrow cells that normally produce platelets (small cells that affect the blood's ability to clot), and a few types of white

blood cells called neutrophils. CML is associated with an abnormality in chromosome 9 called the Philadelphia chromosome. This abnormality occurs when a portion of the genetic material in chromosome 9 is exchanged with a portion of the genetic material in chromosome 22. Ninety-five percent of patients with CML have this genetic alteration. Like CLL, CML is a slowly developing form of leukemia; patients diagnosed with it may have few or no symptoms for months or years before the disease grows worse.

- Acute lymphocytic leukemia (ALL). ALL is the most common type of leukemia in young children and can be rapidly fatal if not treated. In ALL, the patient's bone marrow produces a large number of immature malignant lymphocytes that crowd out healthy blood cells, both red and white. Children with ALL are vulnerable to infection and easy bleeding. The abnormal WBCs can also collect in certain areas of the body, including the central nervous system and spinal cord. This buildup can cause such symptoms as severe headaches, difficulty breathing, a swollen liver and spleen, and dizziness.
- Acute myeloid leukemia (AML). AML is caused by the rapid multiplication of abnormal and immature neutrophils or similar cells that build up within the bone marrow and interfere with the production of normal cells. It worsens quickly if not treated, but it may respond well to therapy, at least in the beginning. Unfortunately, many patients with AML suffer relapses.

# Demographics

There are about 31,000 cases of leukemia diagnosed in the United States each year, 2,000 in children and 29,000 in adults. Of the four major

types of leukemia, about 14,000 cases of CLL are diagnosed each year, almost all of them in adults over fifty-five; 4,400 cases of CML, mostly in adults; 3,800 cases of ALL, almost all in children; and 11,000 cases of AML, which affects both adults and children.

Two-thirds of patients with CLL are men; ALL is slightly more common in men and boys than in women and girls; and about 60 percent of patients with AML are men.

ALL is more common in Italy, the United States, Switzerland, and Costa Rica than in other countries; AML is more common in Caucasians in the United States than in other ethnic groups. CLL is more common in Jewish people of Eastern European descent than in other ethnic groups.

# **Causes and Symptoms**

The causes of leukemia are not completely understood. What is known is that there are several risk factors for these forms of cancer.

- Exposure to high levels of radiation, most often from radiation used to treat other forms of cancer or from nuclear accidents.
- Exposure to certain chemicals, such as benzene or formaldehyde. This type of exposure is most likely to affect adults.
- Chemotherapy for other forms of cancer. Adults treated with certain types of cancer-killing medications may develop leukemia later on.
- Down syndrome. People with this particular genetic disorder have higher rates of leukemia than people in the general population.
- Chromosomal abnormalities such as the Philadelphia chromosome.

It is important to keep in mind, however, that most people with these risk factors do not develop leukemia, and that many people who do suffer from leukemia have none of these risk factors.

The early symptoms of leukemia may develop gradually rather than suddenly and are often mistaken for the symptoms of other diseases. About 20 percent of patients with chronic leukemia do not have any noticeable symptoms at the time they are diagnosed—most often as the result of a routine blood test. Common symptoms of leukemia include:

- Fever and night sweats
- Feeling tired much of the time
- Getting frequent colds and other infections

- Headaches
- Pain in the bones or joints
- Swelling or pain in the abdomen from enlargement of the spleen
- · Cuts or sores taking an unusually long time to heal
- Swelling of the lymph nodes in the neck or armpit
- Unintentional weight loss
- Soft tissue bruising easily, with frequent purple areas or pinpoint bruises under the skin
- Gums and open cuts bleeding easily
- Shortness of breath
- Nausea or vomiting
- In some cases, confusion, dizziness, seizures, or blurred vision

## Diagnosis

The diagnosis of leukemia is complicated by the fact that most of its early symptoms are nonspecific; that is, they occur in many other diseases. The diagnosis is made by a combination of blood tests to check the patient's white blood cell number and kind, followed by a bone marrow biopsy. To do the biopsy, a hematologist (doctor who specializes in the diagnosis and treatment of blood disorders) draws a sample of bone marrow (usually from the hip bone) through a needle after the patient has been given a local anesthetic. The biopsy is necessary to confirm the diagnosis because some diseases other than leukemia can cause an abnormally high number of white blood cells, and some leukemias can only be found in early stages in the bone marrow.

In some cases the doctor will also order a chest x ray or a spinal tap to check for signs of leukemia. In a spinal tap, a small amount of cerebrospinal fluid is removed from the spinal column through a needle. It is done to see whether the disease has spread to the brain or spinal cord.

## Treatment

Treatment for leukemia varies according to the type of disease:

• CLL: Low-grade forms of CLL may not be given any form of treatment because patients do not benefit from therapy in the early stages of the disease. Patients are usually treated when their RBC count or platelet count starts to drop, the lymph nodes become painful, or the number of abnormal WBCs starts to rise sharply. Patients are usually treated with combination chemotherapy; younger patients sometimes benefit from bone marrow transplantation.

- CML: There are several anticancer drugs that can be used to treat CML, but in recent years the standard treatment is a drug called Gleevec, which has relatively few side effects and can be taken by mouth at home. About 90 percent of patients can be maintained on Gleevec for five years without the disease becoming worse. If the drug stops working, bone marrow transplantation is an option; however, the procedure is risky as 30 percent of CML patients die shortly after the operation.
- ALL: Treatment of ALL is focused on preventing the disease from spreading into the central nervous system. It generally has four phases: a beginning phase of chemotherapy to stop the production of abnormal WBCs in the bone marrow; a second phase of medication therapy to eliminate remaining leukemia cells; a third phase of radiation or chemotherapy to prevent the disease from spreading to the brain and spinal cord; and maintenance treatment with chemotherapy to prevent the disease from recurring. ALL can also be treated by bone marrow transplantation.
- AML: AML is treated primarily by chemotherapy in two stages: an induction phase in which the patient is given drugs to reduce the number of cancerous blood cells to an undetectable level; and a second or consolidation phase to eliminate any remaining abnormal cells.

Treatment for leukemia also includes antibiotics to help fight infections when needed, since patients with this type of cancer are vulnerable to infection. The doctor will also provide advice about nutrition and refer the patient to a dietitian if necessary to make sure that the patient is eating a healthy diet and is not losing weight.

Although chemotherapy is the mainstay of treatment for leukemia, in some cases the doctor may recommend surgery to remove the spleen if it has become enlarged. This operation is usually done to control pain and avoid pressure on other organs in the patient's abdomen.

## Prognosis

The prognosis of leukemia depends on the specific type. In general, females have a better prognosis than males.

- CLL: The five-year survival rate is 75 percent.
- CML: The five-year survival rate is 90 percent.
- ALL: Survival rates vary depending on the patient's age. The fiveyear survival rate is 85 percent for children but only 50 percent for adults.
- AML: The five-year survival rate is 40 percent.

#### Prevention

There is no known way to prevent leukemia because the causes of this group of cancers are still not known.

#### The Future

Leukemia is one type of cancer in which survival rates have increased dramatically since the 1960s. In 1960 the overall five-year survival rate for all types of leukemia was about 14 percent; it is over 50 percent as of the early 2000s.

Cancer research in general is a rapidly expanding field. Many new medical centers devoted entirely to cancer research and treatment have been established across the United States. Doctors are testing new approaches to cancer treatment as well as new anticancer drugs, doses, and treatment schedules. Other researchers are working on improving the technique of bone marrow transplantation as a way to treat leukemia, as well as improved methods of diagnosing the disease.

SEE ALSO Down syndrome; Lymphoma

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# WORDS TO KNOW

- Acute: Referring to a disease or symptom that is severe or quickly worsens.
- **B cell:** A type of white blood cell produced in the bone marrow that makes antibodies against viruses.
- **Bone marrow:** The soft spongy tissue inside the long bones of the body where blood cells are formed.
- **Chronic:** Referring to a disease or symptom that goes on for a long time, tends to recur, and usually gets worse slowly.
- Hematologist: A doctor who specializes in diagnosing and treating disorders of the blood.

- **Lymphocyte:** The medical term for white blood cells. A lymphocytic anemia is one that affects the cells in the bone marrow that give rise to white blood cells.
- **Lymphoma:** A type of cancer that affects the lymphatic system.

Myeloid: Relating to bone marrow.

- **Philadelphia chromosome:** A genetic abnormality in chromosome 9 associated with CML. Its name comes from the location of the University of Pennsylvania School of Medicine, where it was discovered in 1960.
- **Platelet:** A type of small blood cell that is important in forming blood clots.

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# Lice Infestation

# Definition

A lice infestation is a condition in which lice are present on a person's scalp, body (or clothes), or pubic area. It is called an infestation rather than an infection because the parasites live on or near the skin and outside of the body rather than in the internal organs. Lice are tiny insects that can spread from one person to another through close contact;

through sharing such personal items as clothing, hats, combs, or hairbrushes; or through lying on a bed, pillow, or carpet that has been in contact with someone with lice.

# Description

The three types of lice that infest humans are somewhat different in size and outward appearance. Head lice are 1-2 millimeters long, white or gray in color, and have flattened abdomens. The female louse lays her nits (eggs) close to the base of a hair shaft and attaches them to it with a sticky, glue-like substance. The glue is what makes it so difficult to remove the nits from the hair shaft.

Body or clothing lice are generally larger than head lice, between 2 and 4 millimeters long. The body louse lives in the seams of clothing, emerging at night to feed on the person's body. Pubic lice are smaller and broader, about 1.2 millimeters long. They have larger front claws, which is why pubic lice are sometimes called "crabs." The claws enable pubic lice to cling to the coarse hairs in the human groin and armpit areas.

# Demographics

The demographics of lice infestation vary depending of the type of lice involved. On the whole, lice infestations are common in the general population. There are at least 12 million cases in the United States each year, although this figure is only an estimate. Head lice infestations are often not reported because people find them socially embarrassing—even though the Centers for Disease Control and Prevention (CDC) states that: "Personal hygiene or cleanliness in the home or school has nothing to do with getting head lice." The number of all three types of lice infestations in North America has increased in recent years.

Head lice are most common in schoolchildren between the ages of three and eleven, and their families. Girls are more likely to be infested than boys because they are more likely to share clothing and other personal items with friends. It can be difficult to prevent a child from picking up head lice at school because of the amount of close contact among children and their belongings.

Body lice infest both children and adults. There is no difference in frequency between men and women. Homeless people and others who live in crowded conditions without opportunities to bathe or shower regularly are at greatest risk of getting body lice. Because the body louse lives in clothing, infestations occur in colder climates. Pubic lice are most

# Also Known As

Pediculosis, crabs

#### Cause

Wingless parasitic insects that live on the head or body, or in the pubic hair

#### Symptoms

Itching; small red bumps on scalp or neck; visible lice or nits (eggs)

#### Duration

Can last from initial infestation until effective treatment; may be recurrent common in people between the ages of fourteen and forty who are sexually active.

There is some seasonal difference in lice infestations in temperate climates. Head lice infestations are more common during the warmer months while body and pubic lice infestations are more common in the fall and winter.

# **Causes and Symptoms**

The cause of lice infestations is the presence of head, body, or pubic lice on a person's body or in his or her clothing. The life cycle of lice helps to explain some of the symptoms of an infestation. When the nit or egg hatches, about eight or ten days after being laid, it produces an immature louse called a nymph. The nymph needs blood to survive. It has sucking mouth parts on its head that can pierce the skin and draw blood to feed on. Human lice must feed about five times a day to survive, otherwise they become dehydrated and die. The nymph becomes a mature adult about ten days after hatching. Its complete life cycle is between thirty and thirtyfive days in length.



The symptoms of a lice infestation depend on the area of the body that is affected:

- Head lice: itchy scalp due to an allergic reaction to the bites of the lice; small red bumps on the head or neck; sensation of something moving over the scalp; an irritated rash caused by scratching the itchy parts of the scalp.
- Body lice: itching and a rash caused by an allergic reaction to the bites of the lice. A long-term infestation may cause discoloration of the skin of the waist area and upper thighs. There may also be open sores caused by scratching the itching areas; these raw areas can become infected by other disease organisms.
- Pubic lice: itching in the genital area or other body areas with coarse hair (armpits, mustache area, eyebrows, eyelashes), and visible nits or lice crawling in the affected area.

*Human hair infested with lice and nits.* ST. BARTHOLOMEW'S HOSPITAL/PHOTO RESEARCHERS, INC.

## A Poem to a Louse

Most people would not think of a head louse as a pleasant topic for a poem. But one of the best-known poems by Robert Burns (1759– 1796), Scotland's national poet, is about seeing a louse crawling up and down a lady's bonnet during a church service. The poem, written in 1786, is a commentary on class snobbery.

In the poem, Burns appears to criticize the louse for its "impudence" in choosing "sae fine a lady" for its host. He tells the creature to "gae somewhere else and seek your dinner/On some poor body." But it is obvious by the end of the poem that he is poking some fun at the well-dressed lady, proud of her expensive bonnet, who is totally unaware that a parasite she would associate with poor people has attached itself to her own head. Burns wrote in Scots dialect rather than standard modern English.

#### Diagnosis

The diagnosis of lice infestation is usually made by examining the skin, hair, pubic area, or clothing of the affected person. The doctor can collect nits from the hair by using a fine-toothed comb or remove lice from the body with a piece of cellulose tape. The organisms can then be studied under the microscope to determine the type of lice involved.

Another test that can be performed involves the use of a Wood's lamp, which is a device that uses ultraviolet light to detect lice, fungal infections, and a few other types of skin infections. The patient is taken into a dark room while the doctor shines the lamp on the area that may be infested. If lice or nits are present, they will glow greenish-yellow.

#### Treatment

Treatment of head lice requires washing the infested person's clothing and bedding in hot

water at 130°F (54.5°C) two days prior to treatment. Clothing that is not washable should be dry-cleaned. Combs and brushes should be soaked in rubbing alcohol for an hour or washed in soap and hot water. These steps are necessary to lower the risk of reinfestation. Toys or other personal items can also be cleared of lice by sealing them inside a plastic bag for at least two weeks; the lice will die from lack of air and food.

The treatment itself consists of applying an over-the-counter or prescription shampoo to the scalp that contains a drug that kills lice. The product directions should be followed exactly regarding how long the product should be left on the hair and whether it should be rinsed out afterward. Following the application, a fine-toothed or special nit comb should be used to comb nits out of the hair. This combing should be repeated every two to three days for three weeks to make sure all the lice and nits are gone. Retreatment with the medicated shampoo may be necessary.

Another treatment that has been found effective in treating head lice is a drug called Mectizan (ivermectin), which was originally

# WORDS TO KNOW

Crabs: A slang term for pubic lice.

**Host:** An organism that is infected by a virus, bacterium, or parasite.

**Infestation:** A condition in which a parasite develops and multiplies on the body of its host rather than inside the body.

Nits: The eggs of lice.

**Wood's lamp:** A special lamp that uses ultraviolet light to detect certain types of skin infections and infestations. It was invented in 1903 by physicist Robert Wood.

developed to treat intestinal parasites. The person needs to take only one dose of the drug.

Body lice are treated by removing all the infested person's clothing and either destroying or washing them in hot water followed by at least twenty minutes in a dryer on the hot setting. The person must take a shower and change into clean clothing. A lice-killing shampoo may be applied to the hairy parts of the person's body.

Pubic lice are treated by applying a shampoo, mousse, or lotion containing a drug called pyrethrin. These products can be purchased over the counter at a pharmacy and should be used exactly as directed on the container. There is also a drug called Ovide that requires a doctor's prescription that can also be used to treat pubic lice. Pubic lice in the eyebrows must be treated by applying a prescription ointment, as Ovide and the over-the-counter products should not be used close to the eyes.

## Prognosis

The prognosis for treatment of lice is extremely good provided care is taken afterward to prevent reinfestation. Head lice and pubic lice are not known to spread other diseases. Body lice, however, are dangerous because they can transmit three potentially fatal illnesses: typhus, relapsing fever, and trench fever.

## Prevention

Practicing good personal hygiene, avoiding sharing personal items with others, and prompt treatment of lice infestations are the best ways of preventing the spread of lice.

#### The Future

Lice are not likely to disappear from the human scene any time soon. Scientists estimate that lice have been infesting humans for at least 3 million years. It is possible that researchers will discover more effective shampoos or soaps to get rid of lice, but for the time being, the best medicine is prevention.

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# m Lung Cancer

# Definition

Lung cancer is the uncontrolled growth of malignant cells in one or both lungs. There are two major types of lung cancer, small cell lung cancer (SCLC) and non-small cell lung cancer (NSCLC). NSCLC is the more common of the two types, accounting for about 87 percent of cases. It develops in the cells of the tissues that line the lungs. SCLC, which is sometimes called oat cell cancer, accounts for the other 13 percent. It develops out of the hormone-producing cells in the lungs and grows more quickly than the NSCLC type of lung cancer. Small cell lung cancer is also more likely to spread to other parts of the body.

Both types of lung cancer may be either primary or secondary. A primary lung cancer is one that starts in the lung and metastasizes (spreads) to other parts of the body—most commonly to the adrenal glands, bones, liver, and brain. A secondary lung cancer is one that began in another organ and spread to the lungs. For example, breast cancer is a type of cancer that frequently spreads to the lungs.

## Description

Lung cancer was a rare disease before smoking tobacco products became widespread; it was not even recognized as a distinct illness until 1761. It is now known to begin when tobacco smoke or some other irritant damages the cells of the lung tissue. The body can repair this damage for some time; eventually, however, the injured cells begin to multiply abnormally, forming a tumor in the lung tissue. The tumor may grow large enough to put pressure on the airway, causing the coughing and difficult breathing that are characteristic of advanced-stage lung cancer.

Another development that can occur is that the cancerous cells in the lung tissue can enter the blood and lymph vessels that supply the lungs. The circulation of the blood and lymphatic fluid can then carry the cancerous cells to other parts of the body. It is possible for the primary lung cancer to metastasize to other organs before coughing or other symptoms appear in the patient's lungs.

Many lung cancers are richly supplied with blood vessels close to the surface of the tumor. If the surface of the tumor is fragile, it may break off and cause bleeding into the airway. The blood may then be coughed up by the patient. Another complication that can develop is pneumonia. If the lung cancer is large enough to partially block the airway, mucus and tissue fluid may build up in the lung tissue behind the blockage, thus making it easier for bacteria to multiply and cause infectious pneumonia.

## Demographics

Lung cancer is the leading cause of cancer deaths worldwide; about 1.3 million people die each year from the disease, 162,000 of them in the

Also Known As Bronchogenic carcinoma

#### Cause

Smoking, exposure to asbestos or radon, or unknown causes

#### Symptoms

Shortness of breath, coughing up blood, chest pain, hoarse voice, wheezing

Duration Years



*Chest x ray of a patient with lung cancer.* © PHOTOTAKE INC. / ALAMY.

United States. Fewer than half of newly diagnosed lung cancer patients live beyond a year after diagnosis; and only 14 percent survive for five years. Lung cancer represents 15 percent of all cancer diagnoses in North America and 29 percent of all cancer deaths.

Lung cancer is a highly preventable disease. Although some risk factors for lung cancer cannot be changed, avoiding tobacco would reduce deaths by about 80 percent. Men who smoke are twenty-three times more likely to develop lung cancer than men who have never smoked; women who smoke have a risk thirteen times greater than that of nonsmokers. In addition to active smoking, the risk factors for lung cancer include:

- Exposure to secondhand tobacco smoke. Nonsmokers who share housing or office space with heavy smokers have an increased risk of lung cancer.
- Exposure to radon. Radon is an invisible, odorless gas produced by the breakdown of uranium in soil and rock. Between 9 and 14 percent of deaths from lung cancer are caused by exposure to radon.
- Occupational exposure to asbestos, uranium, and coke (a fuel used in iron manufacturing).
- Air pollution.
- Age. Lung cancer is almost entirely a disease of older adults. The average age at diagnosis in the United States is seventy years.
- Sex. Men are more likely than women to develop lung cancer; however, the rates for women have risen sharply in recent years because of the increase in smoking among women starting in the 1960s. Nonsmoking women are more likely to develop lung cancer, however, than nonsmoking men.
- Race. African Americans of either sex are more likely to develop and die from lung cancer than any other ethnic group in the United States. On the other hand, Native Americans have one of the lowest rates. The reasons for these differences are not yet known.

- Family history. People with a parent or sibling diagnosed with lung cancer are at increased risk of developing the disease themselves even if they do not smoke.
- Personal history of bronchitis or repeated episodes of pneumonia. Some researchers think that a history of lung disease is a risk factor for eventual lung cancer.

# **Causes and Symptoms**

The largest single cause of lung cancer is exposure to tobacco smoke, followed by such other irritants as radon, asbestos, and air pollution. The causes of lung cancer in nonsmokers are not yet fully understood. Some researchers think that damage to chromosomes 3, 5, 13, and 17 increases a nonsmoker's risk of small cell lung cancer. Another theory concerns human papillomavirus, which has been shown to cause lung cancer in animals. These scientists think that human papillomavirus (HPV, a sexually transmitted virus) infection may trigger lung cancer in some people by causing uncontrolled cell division in lung tissue.

Lung cancer often does not have symptoms in its early stages. A primary lung cancer may produce the following symptoms:

- Fatigue
- Coughing that does not go away
- Coughing up blood
- Chest pain
- Shortness of breath
- Loss of appetite and unintended weight loss
- · Coughing up large quantities of mucus

A lung cancer that has spread to other organs may produce bone pain, abdominal or back pain, headache, weakness, seizures, or speech difficulties.

# Diagnosis

Lung cancers are sometimes diagnosed relatively early when a person develops pneumonia and the doctor discovers a cancerous tumor. In most cases, however, the tumor is diagnosed when the person develops the symptoms of advanced-stage lung cancer.

There is no universally accepted screening test for lung cancer. Some doctors think that a newer type of computed tomography (CT) scan

called a spiral CT scan is a useful way to screen for lung cancer. In a spiral CT scan, the patient lies on a table while the scanner rotates around them. Other doctors, however, think that this test does not yet distinguish clearly enough between lung cancer and other less serious lung problems to justify using it as a screener.

The tests that are most commonly used to detect lung cancer and determine whether it is SCLC or NSCLC include:

- Imaging studies, usually a CT scan of the lungs or an x-ray image of the chest.
- Sputum sample. The patient is asked to cough up some sputum (mucus or phlegm), which can be studied under a microscope for the presence of cancer cells.
- Tissue biopsy. Samples of suspicious tissue may be obtained in one of several ways. The doctor may use an instrument called a bronchoscope (a lighted tube passed down the throat and into the lungs), or make an incision at the base of the neck and remove a tissue sample from the space behind the breastbone. A third technique involves inserting a needle through the chest wall directly into the suspected tumor to remove a sample of tissue.
- Thoracentesis. This is procedure in which the surgeon inserts a needle through the chest wall in order to withdraw some tissue fluid from the space between the lung and the chest wall. As with a sputum sample, the fluid can be checked for cancer cells.

After determining whether the cancer is small cell or non-small cell in type, the next step is staging. Staging is a description of the location of the cancer, its size, how far it has penetrated into healthy tissue, and whether it has spread to other parts of the body. SCLC and NSCLC tumors are staged differently because these two types of lung cancer are treated differently.

- SCLCs are staged in two stages, limited and extensive. A limitedstage SCLC is found only in one lung and its nearby tissues. An extensive tumor is found outside the lung in which it started or in distant organs.
- NSCLCs are staged in an occult (hidden) stage, in which the cancer is detectable only in cells from a sputum sample without

a visible tumor; and five stages graded from 0 to IV in which there is a visible tumor. The grade of the tumor is based on its size and on whether it has spread to the lymph nodes or nearby tissues. In stage 0, for example, the cancer is found only in the innermost lining of the lung. In stage IV, the cancer has spread from one lung to the other lung, or has spread to the brain, bones, liver, or other organs.

#### Treatment

Treatment of lung cancer depends on which type it is and its stage.

SCLCs: Limited-stage small cell lung cancers, which account for about 30 percent of those diagnosed, can usually be treated with radiation therapy. Extensive SCLCs cannot be completely treated with radiation therapy alone and usually require a combination of radiation therapy and chemotherapy.

NSCLCs: Patients diagnosed with non-small cell lung cancers may have surgery, chemotherapy, radiation therapy, or a combination of treatments. The treatment choices are different for each stage. Surgery, for example, may involve removing only a wedge-shaped portion of a lung, an entire lobe of a lung, or the complete lung.

#### Prognosis

The prognosis for lung cancer is poor. It has one of the lowest five-year survival rates of all cancers—about 14 percent as of 2008. For SCLCs, the overall five-year survival rate is 5 percent, with patients diagnosed with extensive disease having a five-year survival rate of less than 1 percent. The average length of survival time for patients with limited-stage disease is 20 months.

For patients with NSCLCs, those with stage I disease treated with surgery have a five-year survival rate of 67 percent; the five-year survival rate of patients with stage IV disease is less than 1 percent.

#### Prevention

There are some preventive measures that people can take to lower their risk of lung cancer:

- Don't smoke or quit smoking.
- Avoid secondhand smoke.

#### WORDS TO KNOW

- **Occult:** The medical term for a cancer that is too small to produce a visible tumor.
- **Radon:** A colorless and odorless gas produced by the breakdown of uranium known to cause lung cancer.
- **Targeted therapy:** A newer type of cancer treatment that uses drugs to target the ways cancer cells divide and reproduce or the ways tumors form their blood supply.

• Have the home tested for radon. The American Lung Association or the local public health authorities can provide information on radon testing.

#### The Future

Lung cancer is likely to be a serious health problem throughout the world as people who started smoking heavily in the 1960s are now getting to the age when lung cancer is usually diagnosed. In the United States, as of 2008 the National Institutes of Health (NIH) is conducting or sponsoring over 2,300 studies related to lung cancer.

SEE ALSO Bronchitis; HPV infection; Pneumonia; Smoking

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# u

# Lupus

## Definition

Lupus, an autoimmune disease, is caused by vascular inflammation (vasculitis) that results in significant damage to a number of different body systems and organs, including the joints, skin, kidneys, heart, lungs, and brain. For this reason it is sometimes called a multisystem disease. Its symptoms vary from patient to patient, ranging from mild conditions that can be managed by medications to life-threatening emergencies.

There are four major types of lupus:

- Systemic lupus erythematosus or SLE. This is the form of the disease most commonly meant by lupus. SLE can occur in childhood but is most common in people between the ages of fifteen and forty-five.
- Discoid lupus. Discoid lupus is a skin disorder in which the patient develops thick raised patches of scaly reddened skin on the face or scalp. A small percentage of patients with discoid lupus later develop SLE.
- Drug-induced lupus. This is a form of lupus triggered by medications. It goes away when the patient stops taking the drugs. Druginduced lupus is more common in men than in women.

#### Also Known As

Systemic lupus erythematosus, SLE

**Cause** Unknown

#### Symptoms

"Butterfly" rash on face, unexplained fever, swollen joints, muscle pain, kidney disease

**Duration** Years



Skin rash on a patient with lupus. © SCOTT CAMAZINE / ALAMY

• Neonatal lupus. This is a rare form of lupus that sometimes occurs in babies born to mothers with SLE, Sjögren syndrome, or no disease at all.

## Description

The causes of lupus, a complex disease, are not understood. It is difficult to diagnose because its symptoms are easy to confuse with those of many other disorders and because there is no symptom profile that applies to all patients with lupus. What is known is that lupus is a chronic inflammatory disease in which the body's immune system turns against its own tissues, producing what are called autoantibodies. Autoantibodies are protein molecules that target the person's own cells, tissues, or organs, causing inflammation and tissue damage. In lupus, the autoantibodies damage the blood vessels in such vital organs as the kidneys.

The inflammation of body tissues in lupus leads to a variety of symptoms that may come and go over time as well as vary from patient

to patient. These include aches and pains in joints and muscles, skin rashes, sensitivity to sunlight, unexplained fever, swollen glands, extreme fatigue, hair loss, mouth ulcers, chest pains, easily bruised tissues, and emotional disorders. Periods when the symptoms are absent or low-key are called remissions, and periods when the symptoms return or increase in severity are called flares.

# Demographics

Lupus is primarily a disease of women of childbearing age. It is rarely diagnosed in children except for the neonatal form. In the United States, lupus affects about one person in every 2,000. According to the Lupus Foundation of America, between 1.5 and 2 million people in the United States may have a form of lupus. The actual number may be higher because the diagnosis is often missed by doctors. Around the world, the rate of lupus varies from country to country, from twelve cases per

100,000 people in Great Britain to thirty-nine per 100,000 in Sweden. In New Zealand, there are fifty cases per 100,000 population among Polynesians, compared with only 14.6 cases per 100,000 among white New Zealanders.

In the United States, lupus is three times more common among African Americans than among Caucasians. It is also more common among Hispanics, Asian Americans, and Native Americans. Like many other autoimmune diseases, lupus strikes women nine times more frequently than men. Among males with lupus, older men are more likely to get the disease than younger men. The fact that women of childbearing age are the group most likely to develop lupus is the reason why some researchers think that female sex hormones may be involved in the disease.

## **Causes and Symptoms**

Researchers believe that lupus is the end result of a combination of genetic, hormonal, and environmental factors. At least ten different genes have been identified that increase a person's risk of developing lupus, and the disease is known to run in families. There is no single lupus gene, however. Other factors that are being studied as possible triggers of lupus include sunlight, stress, certain drugs, and viruses.

# Men and Lupus

Like osteoporosis, breast cancer, and rheumatoid arthritis, lupus is a disease that is largely considered a problem for women. But the 10 percent of lupus patients who are male have some difficulties that female patients do not, precisely because of the gender ratio. According to the Lupus Foundation of America, men with lupus are often worried about being seen as less masculine because of the diagnosis, even though they can still be sexually active and have children. In addition to its feminine image, lupus can cause emotional distress for men by limiting their ability to earn a living or to do chores around the house requiring physical labor. Loss of independence, coupled with such physical changes as hair loss and weight gain, can be a heavy blow to a man's self-esteem.

Researchers are also looking into whether lupus in older men is more severe than in women in the same age groups. Some studies have suggested that men with SLE have a higher risk than women of severe damage to the blood vessels, nerves, and kidneys. Currently male patients with lupus are given the same therapies as women.

The symptoms of lupus may appear in almost any body system:

- Skin: Butterfly-shaped facial rash (also called a malar rash); rash elsewhere on body; ulcers in the mouth, nose, or vagina; loss of hair on head; sensitivity to sun exposure. About 90 percent of patients with lupus have symptoms affecting the skin and hair.
- Bones and muscles: Arthritis, muscle cramps, pains in the hands and wrists.
- Blood: Anemia, low white blood cell count, problems with normal blood clotting, Raynaud's phenomenon (loss of blood flow to fingers and toes due to stress or cold exposure).

- Heart and lungs: Inflammation of the lining of the heart (pericariditis) and lungs (pleuritis). About 50 percent of patients with lupus develop some form of lung disease.
- Nervous system: Seizures, psychotic episodes, memory loss, depression, anxiety. These affect about 15 percent of patients with lupus.
- Kidneys and liver: Kidney disease and eventual kidney failure. About 50 percent of patients with lupus have kidney problems.
- Other: Unexplained fever, weight loss or gain, fatigue.

# Diagnosis

There is no single test that can provide a definitive diagnosis of lupus. The disease is not easy to diagnose because it usually develops slowly over a period of years, its symptoms often come and go, and none of them are unique to lupus. The American College of Rheumatology compiled a list of eleven criteria in 1982 to help distinguish lupus from other diseases. Seven of these are symptoms:

- Butterfly (malar) rash on face
- The raised red patches of discoid lupus
- Skin rash triggered or worsened by sun exposure
- Ulcers in the nose or mouth
- Arthritis
- Inflammation of the tissues lining the inside of the lungs or heart
- Seizures or convulsions in the absence of other causes for these events

The other four criteria are test results:

- Abnormally high levels of protein and red or white blood cell fragments in the urine
- Abnormally low red or white blood cell counts
- Presence of antinuclear antibodies (ANA) and anti-double strand DNA in the blood
- Other positive blood tests that indicate an autoimmune disorder

A person should meet four or more of these criteria for the doctor to suspect lupus. The symptoms do not all have to occur at the same time.

# Treatment

There is no cure for lupus. The symptoms of the disease are managed by medications tailored to the location and the severity of the individual patient's symptoms. The patient's doctor may prescribe drugs from any of the following groups:

- Nonsteroidal anti-inflammatory drugs (NSAIDs). These drugs include aspirin and such non-aspirin pain relievers as Aleve and Motrin. They can be used to treat joint pain, bring down fever, and other inflammatory symptoms of mild lupus.
- Antimalarial drugs. There is no known connection between lupus and malaria; however, some drugs used to treat malaria appear to prevent lupus flares as well as treat symptoms.
- Corticosteroids. This group of drugs includes prednisone, one of the drugs prescribed most frequently to treat lupus. Corticosteroids act to bring down inflammation rapidly. Unfortunately, they also have serious side effects, including weight gain resulting from increased appetite, weakened bones, high blood pressure, damage to the arteries, and an increased risk of infections and diabetes. Doctors try to minimize these side effects by prescribing the lowest dose necessary to control symptoms for the shortest possible time.
- Immunosuppressants. These are drugs that work by reducing the overactivity of the immune system that is involved in lupus. Immunosuppressants are generally given only to patients with severe flares that are damaging organ function, or in order to reduce a patient's dose of corticosteroids.

#### Prognosis

The prognosis for lupus is variable depending on the severity of the symptoms. In general, patients whose kidneys or central nervous systems are affected by the disease have a worse prognosis. Men with lupus have a slightly worse prognosis than women.

The overall life expectancy for patients with lupus has improved since the 1950s, when less than 50 percent of patients were still alive five years after diagnosis. For patients diagnosed with SLE in the United States, Canada, and Europe, 95 percent are alive at five years after diagnosis, 90 percent at ten years, and 78 percent at twenty years. The overall death rate for patients diagnosed with lupus is three times that of the general American population.

## WORDS TO KNOW

Flare: A period of worsened symptoms in lupus.

**Malar rash:** The medical term for the butterflyshaped facial rash found in lupus.

**Neonatal:** The medical term for newborn.

**Raynaud's phenomenon:** Discoloration of the fingers and toes caused by blood vessels going into spasm and decreasing the flow of blood to the affected digits.

**Remission:** A period of decreased or absent lupus symptoms.

#### Prevention

There is no way to prevent lupus because the causes of the disease have not been clearly identified.

#### The Future

Researchers are focusing on new treatments for lupus as well as genetic studies of ethnic groups and families at increased risk of the disease. The Lupus Foundation of America has links to registries for individuals and families with lupus and lupus-related conditions at http://www.lupus.org/webmodules/webarticlesnet/templates/new\_aboutfaq.aspx?articleid=384&zoneid=19. The National Institute of Arthritis and Musculoske-letal and Skin Diseases (NIAMS) is presently conducting studies of lupus in African Americans and Native Americans to look for possible genetic factors associated with the high rates of lupus in these groups.

SEE ALSO Chronic fatigue syndrome; Fibromyalgia; Sjögren syndrome

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# Lyme Disease

## Definition

Lyme disease is an infectious disease caused by a spirochete (spiral-shaped bacterium) transmitted to humans by the bite of an infected deer tick. The alternate name of Lyme disease—borreliosis—comes from the scientific name of the bacterium, *Borrelia burgdorferi*. Lyme disease can be classified as an infectious arthritis because the body's immune response to the bacterium produces inflammation and arthritis-like joint or muscle pain in some people.

Lyme disease is considered a zoonosis, or disease transmitted by animals to humans, as well as an emerging infectious disease. Household pets (cats and dogs) that are allowed outdoors can be infected with Lyme disease as well as humans. Typical symptoms of Lyme disease in animals include joint soreness, limping or lameness, fever, and loss of appetite. Also Known As Borreliosis

#### Cause

Spirochete (spiral-shaped bacterium)

#### Symptoms

Fever, headache, fatigue, rash, joint pains, meningitis

**Duration** Weeks to years



A patient's arm with the bull's eye rash typical of Lyme disease. AP IMAGES.

#### Description

An infectious disease, Lyme disease is caused by a spiral-shaped bacterium that lives inside deer ticks. The tick transmits the disease from one animal to another or from animals to humans when it feeds on their blood. The symptoms of the disease vary from person to person. Not everyone who gets Lyme disease has all the symptoms or has them with equal severity.

The first stage of Lyme disease is often (though not always) marked by a red rash known as erythema chronicum migrans, or EM, at the site of the tick bite. The rash may have a circular or bull's-eye appearance. It occurs in about 80 percent of patients within three to thirty days after the bite. The rash expands over the next few days to cover as much as 12 inches (30 centimeters) of skin. Patients may also have flulike symptoms, including fatigue, chills, low-grade fever, headache, muscle and joint aches, and swollen lymph nodes. In some cases, these flu-like symptoms may be the only indication of Lyme infection.

If the infection is not treated, patients may develop a second stage of symptoms that can

include heart palpitations, fatigue, headaches, temporary paralysis of facial muscles, meningitis, or dizziness.

Some patients experience a third stage of the disease, marked by arthritis-like pain in the joints and muscles, numbress in the arms and legs, loss of memory, and other neurological symptoms.

## Demographics

The Centers for Disease Control and Prevention (CDC) reported 23,305 cases of Lyme disease in the United States in 2005. Most occurred in the coastal Northeast, the Mid-Atlantic States, Wisconsin and Minnesota, and northern California. Most cases of Lyme disease occur in the spring and summer months when ticks are most active and people are spending more time outside. In both the United States and Europe, the age groups most likely to be affected are children between the ages of five and nine years, and adults between fifty and fifty-nine. Among children, boys are

# The Long History of Lyme Disease

Lyme disease is named for Old Lyme, the town in Connecticut where an outbreak of the disease among children in the early 1970s was described by a physician at nearby Yale University. The infectious form of arthritis, however, had been described as far back as the early 1900s in Europe. In 1909, Swedish doctor Arvid Afzelius described a patient with a rash now known as erythema chronicum migrans—or simply erythema migrans (EM)—an early symptom of the disease. By 1934 other European doctors had noted that patients with the strange circular rash eventually developed arthritis-like joint pain and in some cases, psychiatric or neurological symptoms. They had also traced the rash to tick bites.

After World War II, European doctors found that newly developed antibiotics were quite effective

in treating the tick-borne disease. The first known case of EM in the United States was reported in 1970 by a doctor in Wisconsin who was treating a patient bitten by a tick while hunting. The first cluster of cases of Lyme disease in the United States occurred in 1976 at a U.S. naval base in Connecticut. The following year saw cases involving the school children of Old Lyme. At that time, the disease was called Lyme arthritis. In 1982 Willy Burgdorfer, a researcher with the Rocky Mountain Laboratories of the National Institutes of Health, identified the cause of Lyme disease while gathering black-legged ticks in Montana. The organism was named Borrelia burgdorferi in his honor. Researchers have shown two other Borrelia species cause the European form of Lyme disease.



Tick capable of carrying the bacteria that causes Lyme disease. SHUTTERSTOCK.

more likely to be infected than girls, but in the older age group, women are slightly more likely than men to get Lyme disease. In 1998, Caucasians accounted for 76 percent of reported cases of Lyme disease in the United States, but it is not known whether this statistic indicates greater susceptibility to the disease or simply regional differences in reporting.

#### **Causes and Symptoms**

The cause of Lyme disease is a spirochete carried from one animal or human host to another by several varieties of ticks found in the United States. These ticks have a two-year life cycle. They are born in the summer as larvae and feed only once, on the blood of field mice. The next spring, the larva becomes a nymph and feeds again on a mouse's blood. In the fall, the nymph becomes an adult tick and feeds on the blood of a white-tailed deer. If the tick has picked up the spirochete from the mice or the deer, it can transmit the disease to a human at this point.

It takes one to three days for the tick to transmit *B. burgdorferi* to a human because it takes time for the bacterium to multiply inside the tick after it has bitten a person. Once feeding begins, the bacteria inside the tick multiply rapidly and move into the salivary glands of the tick after one to two days or so. The tick then injects the bacteria into the human as it continues its feeding. This time delay is one reason why prompt removal of a tick is usually effective in preventing Lyme disease and most other tick-borne infections.

#### Diagnosis

The diagnosis of Lyme disease is complicated by several factors. The first is that only 20 percent of patients are aware that they have been bitten by a tick. If they do not develop the characteristic EM rash, the diagnosis may be delayed. Second, the ticks that carry Lyme disease also often carry other diseases like ehrlichiosis or babesiosis, so that a person may have another tick-borne infection alongside or instead of Lyme disease. Third, most of the symptoms of Lyme disease can be caused by a variety of other disorders, including rheumatoid arthritis, complications of gonorrhea, lupus, or gout.

The CDC recommends as of 2007 that doctors look for three factors when evaluating a patient who might have Lyme disease:

• A history of possible exposure to ticks in parts of the United States known to have a higher than average rate of Lyme disease

- Physical symptoms that include EM
- A blood test that shows the patient has antibodies to B. burgdorferi

Even so, blood tests are not 100 percent accurate, particularly if they are given before the patient's body has had time to develop antibodies to the spirochete. In most parts of the United States, an initial blood test for antibodies is followed up by a second test known as a Western blot test to confirm the diagnosis.

# Diagnosis

Early-stage Lyme disease can be effectively treated with a fourteen- to twenty-one-day course of antibiotics taken by mouth. These drugs usually clear the infection and reduce the risk of later complications. Second- or third-stage Lyme disease is treated with either a thirty-day course of an oral antibiotic or fourteen to twenty-eight days of an intravenous antibiotic.

## Prognosis

The prognosis of Lyme disease is difficult to estimate because of the fact that EM is sometimes misdiagnosed. Further, many patients do not return for follow-up visits, which complicates the doctor's ability to measure the effectiveness of treatment or record the length of time that the patient had symptoms. In general, children who are treated early with antibiotics have an excellent prognosis for complete recovery. Adults are more likely to develop chronic muscle and joint pain or fatigue, but generally recover given time and appropriate treatment. Although there have been a few fatal cases of Lyme disease in humans as of 2008, the overall mortality rate is extremely low.

## Prevention

A vaccine effective against Lyme disease was released in 1998 but was taken off the market because of the possible side effects reported by some patients and because it was not widely used. Although research into a better vaccine is ongoing, there was no vaccine available against the disease as of mid-2008. Preventive measures against Lyme disease are important because of the lack of an effective vaccine. The CDC recommends the following precautions:

• Stay away from wooded, brushy, and grassy areas, especially in May, June, and July. These are the months when ticks are most likely to feed on humans and pets.

- Wear light-colored clothing (which allows the ticks to be seen more easily); shoes that cover the entire foot; long pants tucked into socks or shoes; and long-sleeved shirts tucked into pants. Also wear a hat for additional protection.
- Use insect repellent containing a chemical called DEET on clothes and exposed skin other than the face. Another repellent that can be used on clothing is permethrin, which kills ticks on contact.
- When hiking in the woods, walk in the center of the trail or path; avoid brush and tall grasses.
- Remove clothing after being outside; wash it in hot water and dry it in a dryer on a high setting.
- Check the body for ticks after being outside, particularly the hair, the groin area, and the armpits.
- Check pets allowed outdoors for ticks in their fur, and give them tick-repellent collars. There is a vaccine to protect dogs against Lyme disease; there is, however, no vaccine for cats as of 2008. Cats appear to be much less likely to get the disease than dogs, which can develop fatal kidney problems from Lyme disease.
- Remove a tick properly if one is found on the body. Grasp the tick near its head or mouth, as it is critical to remove the head intact. Do not crush the tick but pull it backward from the skin slowly and carefully. Take the tick to a doctor or local public health department so that it can be tested to see if it is a Lyme diseaserelated tick.

## The Future

Lyme disease is likely to become more common in North America in the years ahead because of the rising number of deer, mice, and other small rodents that can be infected by *B. burgdorferi*, and the increased amount of contact between humans and these animals in wooded areas. This increased contact is partly due to the growing popularity of woodland hiking and fishing and partly to the building of new houses in tick-infested areas. The CDC reports that Lyme disease was one of the fastest-growing infectious diseases in the United States as of 2008 and that it has spread from the regions where it was first noticed to forty-nine of the fifty states. Researchers are working on developing a new vaccine against Lyme disease. In addition, other scientists are trying to learn more

# WORDS TO KNOW

- **Babesiosis:** A malaria-like disease that can be transmitted by ticks.
- **Ehrlichiosis:** A tick-borne disease found primarily in dogs that can also be transmitted to humans.
- **Emerging infectious disease (EID):** A disease that has become more widespread around the world in the last twenty years and is expected to become more common in the future.
- **Endemic:** A term applied to a disease that maintains itself in a particular area without reinforcement from outside sources of infection.
- **Erythema chronicum migrans (EM):** The medical name for the distinctive rash that is often seen in early-stage Lyme disease.

- **Larva:** The immature form of a deer tick.
- **Meningitis:** Inflammation of the membranes covering the brain and spinal cord.
- **Nymph:** The second stage in the life cycle of the deer tick.
- **Spirochete:** A spiral-shaped bacterium. Lyme disease is caused by a spirochete.
- **Tick:** A small bloodsucking parasitic insect that carries Lyme disease and several other diseases.
- **Zoonosis:** A disease that animals can transmit to humans.

about the bacterium that causes the disease in order to develop better ways to diagnose and treat it.

SEE ALSO Lupus; Rheumatoid arthritis

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# Lymphoma

# Definition

Lymphoma refers to a varied group of cancers of the blood that develop from white blood cells in the lymphatic system. The lymphatic system is a group of organs and tissues that are part of the immune system and also help to form new blood cells. It includes lymph nodes, small organs composed of lymphoid tissue located at various points throughout the body that are joined by lymphatic vessels; the spleen, a small organ on the left side of the abdomen that produces lymphocytes and stores red blood cells; the bone marrow, which produces new red and white blood cells; and the thymus gland just below the neck, which produces one type of lymphocyte, the T cell.

Two major types of lymphoma were defined in the early 1980s— Hodgkin disease, sometimes called Hodgkin lymphoma or HL; and non-Hodgkin lymphoma or NHL. HL was named for Thomas Hodgkin (1798–1866), a British doctor who first described it in 1832, and was the first form of lymphoma to be officially defined, in 1963. Researchers focused on Hodgkin disease relatively early because it can be treated effectively by radiation therapy. Other forms of lymphoma were then grouped under the general heading of non-Hodgkin lymphoma or NHL in 1982. This entry will focus on non-Hodgkin lymphoma or NHL.

# Description

Like Hodgkin disease, non-Hodgkin lymphoma begins in the lymphocytes, or white blood cells in the immune system. About 85 percent of

#### Also Known As

Lymphocytic lymphoma, non-Hodgkin lymphoma, NHL

**Cause** Unknown

#### Symptoms

Swollen lymph nodes, night sweats, fever, weight loss, lack of energy

#### Duration

Lifelong unless treated

#### Lymphoma

NHLs originate in B cells, which are lymphocytes produced in the bone marrow. Most of the remaining 15 percent develop from T cells produced in the thymus gland. What happens is that the abnormal B or T cells start to multiply uncontrollably, often within the lymph nodes, causing swelling and pain. The lymphoma can spread from the lymph nodes to the lymphatic vessels, tonsils, adenoids, spleen, thymus, and bone marrow. A non-Hodgkin lymphoma can also spread outside the lymphatic system to such other organs as the liver.

Non-Hodgkin lymphomas vary considerably in their speed of development and danger to survival. The 1982 classification categorized NHLs as low-grade, intermediate-grade, or highgrade depending on their aggressiveness and the organs affected by the cancer. Low-grade lymphomas are sometimes called indolent lym-



phomas because they grow slowly and cause relatively few symptoms. Intermediate-grade and high-grade lymphomas grow and spread more rapidly and cause severe symptoms. *Image of lymphoma cells grown in a laboratory.* STEVE GSCHMEISSNER / PHOTO RESEARCHERS, INC.

#### **Demographics**

Non-Hodgkin lymphoma accounts for about 4 percent of all cancer diagnoses in the United States. It is seventh in frequency among all cancers and is five times more common than Hodgkin disease. About 64,000 Americans are diagnosed with NHLs each year, and 18,700 die from this form of cancer. The five-year survival rate for non-Hodgkin lymphomas is 63 percent.

Non-Hodgkin lymphomas usually affect older adults; they are most likely to occur in people over sixty. Low-grade lymphomas account for 37 percent of NHLs in patients aged thirty-five to sixty-four but account for only 16 percent of cases in patients younger than thirty-five. Lowgrade lymphomas are extremely rare in children.

Men are slightly more likely than women to develop non-Hodgkin lymphomas; the gender ratio is 1.4:1. Caucasians are more likely to develop NHLs than either African Americans or Asian Americans. Risk factors (other than age) for non-Hodgkin lymphoma include:

- Exposure to certain chemicals, particularly benzene and certain weed-killing chemicals
- Exposure to radiation, including nuclear reactor accidents as well as radiation treatment for cancer
- Taking drugs that suppress the immune system, including chemotherapy for cancer as well as drugs given to prevent rejection of a transplanted organ
- Certain infections, including AIDS and infection with a bacterium associated with stomach ulcers
- Autoimmune diseases, including lupus and rheumatoid arthritis
- Extreme obesity

# **Causes and Symptoms**

Non-Hodgkin lymphomas are caused by the uncontrolled multiplication of abnormal B or T cells. What triggers the formation of the abnormal cells is not completely understood but is thought to be related to the activation of abnormal genes called oncogenes. Oncogenes are genes that have the potential to trigger normal cells into becoming cancerous.

The most common symptoms of NHLs are:

- Swollen but painless lymph nodes in the neck, groin, or armpit areas
- Fever
- Unexplained or unintended weight loss
- Soaking night sweats
- Coughing or difficulty breathing
- Chest pain
- Fatigue that does not go away
- Itchy skin
- Pain, swelling, or a feeling of fullness in the abdomen

A few patients may have no symptoms at all in the early stages of the disease other than swollen lymph nodes.

## Diagnosis

The diagnosis of NHLs can be complicated because none of the symptoms of lymphomas are unique to this type of cancer. The first step in diagnosing non-Hodgkin lymphoma is to rule out other diseases that can cause swollen lymph nodes. In addition to examining the patient's lymph nodes as part of a physical examination, the doctor will order blood and urine tests to see whether an infection might be the cause of the patient's symptoms. The doctor will also ask how long the symptoms have been present; while the flu can cause fever and fatigue, for example, those symptoms should go away after a week or two.

The next step in diagnosis is imaging studies, including a chest x ray and a computed tomography (CT) scan or magnetic resonance imaging (MRI) study of the chest, abdomen, or pelvic area. These tests can identify the location and size of tumors within the lymph nodes in those parts of the body. A newer type of imaging test that may be ordered to detect lymphoma is a positron emission tomography (PET) scan. In a PET scan, a radioactive substance called a tracer is injected into the patient's circulation. The radioactive material tends to concentrate in tissues that show an increased level of metabolic activity, which often means a tumor.

In addition to imaging studies, the doctor will collect a tissue sample called a biopsy to be examined under the microscope in a specialized laboratory. If the swollen lymph node is close to the surface of the skin, the doctor can remove the tissue sample through a hollow needle. If the lymph node lies deeper within the body, a surgeon may be called in to remove the tissue by making an incision.

To determine whether the lymphoma has spread, the doctor may also order a bone marrow biopsy. In this test, the patient is given a local anesthetic and a sample of bone marrow is removed from the hip bone through a hollow needle.

#### Treatment

The first step in treating any kind of cancer is called staging. Staging is a description of the location of the cancer, its size, how far it has penetrated into healthy tissue, and whether it has spread to other parts of the body. Non-Hodgkin's lymphoma is classified into four stages:

- Stage I: The disease is limited to one lymph node group or one tissue or organ (such as the spleen or liver).
- Stage II. The disease involves two or more lymph node groups on the same side of the diaphragm, or in one part of an organ and the lymph nodes near that organ.

- Stage III. The disease has spread to lymph node groups on both sides of the diaphragm and may involve a part of an organ or tissue near those groups.
- Stage IV. The disease has spread to several parts of one or more organs in addition to the lymph nodes.

The treatment of non-Hodgkin lymphoma depends on the subtype to which the tumor belongs and its stage of development.

- If the patient has an indolent NHL without symptoms, the doctor may recommend watchful waiting rather than beginning treatment right away, as all forms of cancer therapy have some side effects.
- Early-stage NHLs are treated with either radiation therapy or a combination of radiation and chemotherapy. Chemotherapy for lymphoma usually involves a combination of drugs rather than a single agent. It may be given either intravenously or by mouth.
- Early-stage lymphomas may also be treated with biological therapy, which involves vaccines and other drugs intended to boost the functioning of the patient's immune system. Biological therapy is also given to offset some of the side effects of radiation and chemotherapy.
- Aggressive lymphomas are treated with a combination of chemotherapy and biological therapy.
- Patients whose cancers return after therapy are given high doses of radiation, chemotherapy, or both, followed by stem cell transplantation. This procedure involves giving the patient stem cells after chemotherapy in order to help the patient's bone marrow recover and begin to produce healthy blood cells again.

## Prognosis

The prognosis of non-Hodgkin lymphoma depends on the specific tumor type and location; the patient's age; severity of symptoms; the patient's ability to tolerate intensive chemotherapy; and whether the disease has spread beyond the lymph nodes. In general, patients older than sixty, patients with weakened immune systems, and patients with T-cell lymphomas have worse prognoses than younger patients, patients who are otherwise healthy, and patients with B-cell lymphomas.

About 70 percent of patients with intermediate- or high-grade lymphomas at the time of diagnosis either fail to respond to treatment or

# WORDS TO KNOW

- **Biological therapy:** An approach to cancer treatment that is intended to strengthen the patient's own immune system rather than attack the cancer cells directly.
- **Diaphragm:** A sheet of muscle extending across the bottom of the rib cage that separates the chest from the abdomen.
- **Indolent:** The medical term for a tumor or disease that grows or develops slowly.
- **Lymph nodes:** Small rounded masses of lymphoid tissue found at various points along the lymphatic vessels.
- **Lymphocyte:** A type of white blood cell that fights infection. Lymphocytes are divided into two

types, T cells (produced in the thymus gland) and B cells (produced in the bone marrow).

- **Oncogene:** A gene that has the potential to cause a normal cell to become cancerous.
- **Staging:** Measuring the severity or spread of a cancer.
- **Stem cell:** A type of body cell that has the ability to differentiate into various types of specialized cells.
- **Thymus:** A small organ located behind the breastbone that is part of the lymphatic system and produces T cells.

have a recurrence of their cancer. About 5 percent of patients with recurrent cancer will survive for two years after the recurrence.

## Prevention

There is no way to prevent non-Hodgkin lymphoma because its causes are still unknown, and some potential risk factors may not yet have been identified.

# The Future

Researchers are looking for an explanation for the rise in the number of cases of NHLs in the United States since the 1970s. The figure nearly doubled between the 1970s and early 2000s. Although some of the increase can be explained by improved diagnostic techniques, there appear to be other factors involved that have not yet been identified.

Other scientists are studying various innovative treatments for non-Hodgkin lymphoma, including new anticancer drugs, new types of biological therapy, and improved methods of stem cell transplantation.

SEE ALSO Hodgkin disease; Leukemia
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# M



## Genetic

Infection

Injury

Multiple

Other

Unknown

## Mad Cow Disease

See Creutzfeldt-Jakob disease.



## Malaria

## Definition

Malaria is an infectious disease caused by a parasite that spends part of its life cycle in humans and part in mosquitoes.

## Description

Malaria is a potentially fatal disease that has infected humans as far back as 50,000 BCE. There are records of mosquito netting being used in Egypt in 2700 BCE to protect against malaria. Chinese medical records from the same period describe the symptoms of the disease. The English name of the disease comes from an eighteenth-century Italian doctor who wrote a textbook about it and attributed it to bad air, or *mal'aria* in Italian.

Humans develop malaria when they are infected with a protozoan called *Plasmodium* through the bite of a mosquito. The parasites enter the bloodstream and are carried to the liver, where they infect liver cells and multiply. The mature parasites are released back into the bloodstream, where they infect red blood cells. The red cells burst in two to three days, releasing more parasites that, in turn, invade more red blood

#### Malaria



Magnified image of a red blood cell infected with the malaria parasite. © PHOTOTAKE INC. / ALAMY.

#### Also Known As

Jungle fever, blackwater fever, *Plasmodium* infection

#### Cause

Parasite carried by a type of mosquito

#### Symptoms

Chills, fever, sweating, joint pains, jaundice, headache, nausea, vomiting, coma, convulsions

#### Duration

May kill within two weeks or develop months to years after exposure cells. Most of the symptoms of malaria (fever, chills, generally sick or flu-like feeling) are related to the destruction of red blood cells. The reason why the fever associated with malaria comes in two- or three-day cycles in some people is that the bursting of infected red blood cells and the infection of new red blood cells occurs every two to three days.

There are four different species of *Plasmodium* that infect people. One of these, *Plasmodium* falciparum, is much deadlier than the

other three. It can cause failure of the lungs, kidneys, and central nervous system within a few hours or days. The different species also take different lengths of time to produce the first symptoms of malaria in humans. Some remain in the liver for long periods of time, thus causing flare-ups of the disease months or even years later. These variations are one reason why doctors try to identify the particular *Plasmodium* species when they test a person's blood for malaria. It is possible for a person to be infected with more than one species of the parasite at the same time.

People cannot get malaria from sharing a household with an infected person since it takes more than a week before a mosquito is able to transmit the disease. It is possible for a pregnant woman to transmit the disease to her unborn baby, however. It is also possible to get malaria through a transfusion of infected blood, but this form of transmission is rare in developed countries. In the United States, people who have traveled in an area with endemic malaria cannot donate blood for a full year after returning to the United States. They are forbidden to donate blood for three years if they have been treated for malaria.

## Demographics

Malaria is one of the most common infectious diseases in tropical and subtropical parts of the world. These countries account for 41 percent of the world's population. It is estimated that there are between 400 and 900 million cases of malaria around the world each year and 1 million to 3 million deaths, most of them African children under age five. In some parts of Africa, it is estimated that one person dies of malaria every thirty seconds. The disease is endemic in parts of Africa, southern Asia, and portions of South America.

## Malaria Research and the Nobel Prize

One indication of the importance—and the complexity—of malaria is the number of Nobel prizes in medicine that have been awarded to doctors who studied the disease. Charles Louis Alphonse Laveran (1845–1922) was a French doctor working in a military hospital in Algeria in 1880 when he discovered that malaria is caused by a protozoan (a type of one-celled organism). This was the first time that a protozoan had been shown to cause any disease in humans. Laveran was awarded the Nobel Prize in Physiology or Medicine in 1907.

In 1898, Sir Ronald Ross (1857–1932), a British doctor working in India, was able to show that mosquitoes are the vectors that carry malaria to humans. Ross even demonstrated that the parasites are present in the mosquito's salivary glands.

He was awarded the Nobel Prize in Physiology or Medicine in 1902. Austrian doctor Julius Wagner-Jauregg (1857–1940) won a Nobel prize in 1927. He showed that malaria could actually be used to treat patients suffering from late-stage syphilis, a common cause of dementia in the early twentieth century. The reason this treatment was thought to work was that the fever caused by the malaria parasite was high enough to kill the bacteria that cause syphilis.

The fourth Nobel Prize in Physiology or Medicine related to malaria research was given in 1948 to Paul Hermann Müller (1899–1965), a Swiss chemist who discovered dichloro-diphenyltrichloroethane (DDT) in 1939. DDT was the first insecticide that was effective in controlling the mosquitoes that spread malaria.



Ronald Ross with his wife on the steps of his hospital laboratory in Calcutta, India.

Malaria has not been endemic in the United States since 1951, when a four-year program was started in 1947 to spray areas in the southeastern part of the country that were known to harbor the mosquitoes that transmit malaria. By 1952 the Centers for Disease Control and Prevention (CDC) reported that malaria had been eradicated from the United States. Cases of malaria since that time were either acquired by people traveling to countries where malaria is endemic or were small local outbreaks caused by local mosquitoes biting infected travelers and then transmitting the malaria parasites to others nearby. In 2002 the CDC reported 1,337 cases of malaria in the United States, including eight deaths. All but five cases were acquired abroad.

People who are at greatest risk for malaria are those who have never been exposed to the disease, pregnant women, and young children. Some researchers also think that genetic factors may increase some people's risk of getting the disease.

#### **Causes and Symptoms**

The cause of malaria is a protozoan of the genus *Plasmodium*. The most deadly of the four species that can cause malaria in humans is *Plasmo-dium falciparum*. The classical symptom of malaria is a repeated cycle of chills and shaking followed by fever and sweating lasting four to six hours. People infected by *P. falciparum* may feel a tingling in the skin as well. In some cases, the early symptoms of malaria may be mistaken for the flu.

Children infected by *P. falciparum* may develop increased fluid pressure on the brain and suffer permanent brain damage even if they survive. Adults infected by *P. falciparum* may develop kidney failure, dehydration, an enlarged liver and spleen, rupture of the spleen, severe headache, loss of blood supply to the brain, and coma. Some die. The destruction of red blood cells by the parasite may cause the urine to look brown or black, which is why the disease got the name "blackwater fever."

#### Diagnosis

Diagnosis of malaria is based on a combination of the patient's history and the results of blood tests. The patient's history is critical, particularly the dates of any trips abroad, because two of the four species of the malaria parasite can remain dormant in the liver for months or even years.

The basic blood test involves looking at blood smears made by pricking the patient's finger at twelve- to twenty-four-hour intervals. One technique is called a thick smear, which is used to estimate the number of parasites in the blood. The other is called a thin smear and is used to determine the species. A rapid test called OptiMAL has been developed to distinguish *P. falciparum* from the other species. It gives results in less time than the smear method and can be used by doctors in areas without hospital laboratories.

## Treatment

Malaria is treated with various combinations of drugs, which may be given by mouth or intravenously. Patients infected by *P. falciparum* may be taken to a hospital for treatment because of the possibility of major organ damage. The oldest drugs used to prevent or treat malaria are quinine and chloroquine. Relatively inexpensive, they are not always effective because the malaria parasites have developed resistance to them in some countries. Newer drugs have been developed that are more effective for resistant strains. People who are traveling to parts of the world where malaria is endemic need to take these drugs while they are abroad.

The specific drug that is given to a patient and the length of treatment depend on the type of malaria, the source of the infection, the patient's age, and the severity of the symptoms when treatment began. People often feel weak and tired for a few weeks when taking these medications.

## Prognosis

Malaria caused by *P. falciparum* is the most deadly; it can kill within days of the first symptoms. In some areas, the death rate from this type of malaria is as high as 20 percent. People infected by the other three species of *Plasmodium* usually recover, but some may have bouts of malaria for months until all the parasites have been cleared from their livers.

### Prevention

There is no vaccine effective against the malaria parasite, although scientists are working on several possibilities. For the time being, prevention is the most effective way to lower one's risk of getting malaria. There are three basic strategies recommended by the CDC:

- Controlling the mosquito population by using insecticides and draining swampy areas. This approach is called vector control.
- Using mosquito netting treated with insecticide to cover beds and other sleeping areas. These nets have cut the number of deaths from malaria in parts of Africa by 20 percent.
- Taking medications to prevent the disease when visiting countries where malaria is endemic. The CDC has general advice for travelers and a list of countries where travelers are at risk for malaria at http://www.cdc.gov/malaria/travel/index.htm.

## The Future

It is unlikely that the mosquitoes that carry malaria will ever be completely wiped out. At present, efforts to relieve the impact of malaria in Africa are aimed at preventing the disease. Methods include spraying insecticides, providing people with insecticide-treated mosquito netting, and supplying poor countries with antimalarial drugs. In 2005 the President's Malaria Initiative (PMI)—a cooperative program that involves the CDC, the National Institutes of Health, the World Health Organization, and the World Bank as well as the White House—began a five-year program to cut deaths from malaria by 50 percent in fifteen African countries.

The most promising area of malaria research as of 2008 was the development of vaccines against the disease. Scientists have identified the genetic sequence of the malaria parasite, which may help researchers target a particular stage in the life cycle of the parasite or prevent it from developing inside the mosquito. One possibility is developing a vaccine that would stop the parasite inside the human body before it gets to the point of infecting the red blood cells. Another group of researchers is experimenting with cultivating *P. falciparum* inside mosquitoes and treating the parasite with radiation in order to make a vaccine. This approach is described in the video listed below.

SEE ALSO Influenza; Syphilis; West Nile virus infection

## For more information

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## WORDS TO KNOW

**Dementia:** Loss of memory and other mental functions related to thinking or problem-solving.

- **Endemic:** A term applied to a disease that maintains itself in a particular area without reinforcement from outside sources of infection.
- **Protozoan (plural, protozoa):** A one-celled animallike organism with a central nucleus enclosed by

a membrane. Many protozoa are parasites that can cause disease in humans.

- **Spleen:** An organ located behind the stomach that cleans old blood cells out of the blood and holds a reserve of red blood cells.
- **Vector:** An insect or other animal that carries a disease from one host to another.

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## Marfan Syndrome

## Definition

Marfan syndrome, or MFS, is a disorder of the connective tissue caused by a mutation in the FBN1 gene on chromosome 15. Because connective tissue occurs throughout the human body, Marfan syndrome affects the patient's eyes, circulatory system, skin, and lungs as well as the bones and muscles. The disorder is named for Antoine Marfan (1858–1942), a French pediatrician who first described it in a five-year-old girl. The gene responsible for MFS was identified in 1991 by Francesco Ramirez, a researcher at Mount Sinai Medical Center in New York.

## Description

Marfan syndrome is a hereditary connective tissue disorder caused by mutations in the FBN1 gene. This gene is responsible for the production of elastic fibers in connective tissue and for the proper timing of the release of growth factors. People with defective FBN1 genes are not affected with equal severity, however; at least 137 different mutations of the gene have been identified. Some people may have only a few of the characteristic features of MFS, while at the other extreme, some babies are born with a severe form of the syndrome that progresses rapidly to early death from heart problems.

The classic signs of Marfan syndrome are long, thin arms and legs, bone overgrowth, and loose joints or poor muscle tone. The most common symptoms of MFS, however, are eye disorders, such as an increased risk of cataracts, dislocation of the lens of the eye, nearsightedness, and glaucoma. These affect more than 50 percent of patients. Skeletal deformities are another common characteristic, particularly scoliosis (abnormal curvature of the spine) and deformities of the ribs and breastbone.

The most damaging health problems associated with Marfan syndrome, however, are those involving the heart and circulatory system. The weakness of the connective tissue in patients with MFS leads to damaged heart valves, a weakened aorta (the large blood vessel that carries blood away from the heart), enlargement of the pulmonary artery (which carries blood from the heart to the lungs), or an aortic dissection.

Also Known As MFS

#### Cause

Mutation in the FBN1 gene on chromosome 15

#### Symptoms

Unusual height, long, thin arms and feet, heart problems, vision problems, poor muscle tone

#### Duration

Lifelong

An aortic dissection is a medical emergency that occurs when a tear in the wall of the aorta allows blood being pumped from the heart under pressure to force its way between the layers of tissue that form the aorta. The separation of the tissue layers can lead to a complete rupture of the aorta and rapid death.

## **Demographics**

MFS affects about one in every 5,000 persons in the United States. It occurs with equal frequency in both sexes and in all races and ethnic groups. It is estimated that as many as 200,000 people in the United States have some form of Marfan syndrome. Many cases are not diagnosed, however, because the affected person may have only mild symptoms of the disorder.

Marfan syndrome is inherited in what geneticists call an autosomal dominant pattern. That means that only one copy of a defective FBN1 gene is needed to produce the condition. Although Marfan syndrome does run in families, between 25 and 30 percent of diagnosed cases involve new mutations of the FBN1 gene in patients with no family history of MFS.



Boy with Marfan syndrome (center) playing basketball. He wears a special vest to protect his heart. AP IMAGES.

Marfan syndrome appears to be equally common in all countries around the world.

## **Causes and Symptoms**

The cause of Marfan syndrome is a mutation in the FBN1 gene on chromosome 15. There are nearly 140 mutations of this gene that have been identified as of 2008. The gene governs the production of a protein called fibrillin-1, which helps to form tiny threadlike filaments that become part of the fibers in connective tissue. The tiny filaments also control the release of growth factors, which are protein molecules that stimulate the growth and multiplication of cells. In normal circumstances, the filaments release these growth factors at the proper time; but in Marfan syndrome, the filaments release the growth factors too soon. The

## Did Abraham Lincoln have Marfan Syndrome?

In 1959, a doctor named Harold Schwartz advanced the theory that Abraham Lincoln suffered from Marfan syndrome. Lincoln had some of the physical features associated with MFS: he was tall (6 feet 4 inches [2 meters]) and thin (160–170 pounds [73–77 kilograms]). Lincoln was also loosejointed, shuffled when he walked, and had to wear eyeglasses. Dr. Schwartz then discovered that Lincoln shared a great-great-grandfather with a man known to have Marfan syndrome, which seemed to confirm the diagnosis.

However, Lincoln had no other signs of the disorder. His visual problem was not myopia, but farsightedness. In addition, he did not suffer from the chest pains, fatigue, and lung problems typical of MFS. In 2007 a cardiologist named John Sotos published a book in which he speculates that Lincoln actually suffered from something much less common, but much more serious than Marfan syndrome—a type of hereditary cancer called multiple endocrine neoplasia type 2B, or MEN 2B. People with MEN 2B develop cancers in hormoneproducing organs like the thyroid or the adrenal gland. Sotos thinks that Lincoln's mother and two of his sons may have also died from MEN 2B.

The historical riddle could be solved by a DNA test. It is known that Lincoln's doctor kept a few locks



*President Abraham Lincoln.* COURTESY OF THE LIBRARY OF CONGRESS.

of his hair and some bloodstained clothing after his assassination, and that the federal government has stored eight fragments from Lincoln's skull. It is not known, however, whether those in charge of these remains would be willing to give up a sample for DNA analysis.

defective fibrillin-1 produced by a mutated FBN1 gene will thus result not only in weakened connective tissue but also in the unusual height and long arms characteristic of MFS patients.

The symptoms in organ systems most commonly affected by MFS are:

• Visual: High risk of cataracts (clouding of the lens), glaucoma (high fluid pressure inside the eye), myopia (nearsightedness), and dislocation of the lens of the eye.

- Circulatory: High risk of widening (resulting in an aneurysm) or dissection of the aorta; defects in heart valves leading to heart murmurs, shortness of breath, tiring easily, chest pain, and a very fast or irregular heart rate.
- Skeletal: Overgrowth of the long bones of the body; loose joints; abnormally shaped mouth with crowded teeth; scoliosis (curvature of the spine); breastbone that either curves inward or protrudes outward; flat feet; long, narrow skull.
- Nervous system: Weakening of the membrane that covers the spinal cord. This defect can lead to pain in the abdomen or legs in middle age.

Other symptoms that some patients with MFS experience include:

- High risk of sleep apnea
- Speech problems caused by the high arch in the roof of the mouth
- Stretch marks in the skin that are not due to weight loss or pregnancy
- Learning disabilities
- Headaches
- Cold hands or feet

## Diagnosis

The diagnosis of Marfan syndrome is complicated for several reasons. One is that there is no single genetic test that can identify all the known mutations of the FBN1 gene. Another is that some of the symptoms of MFS are also found in other connective tissue disorders. A third reason is that some symptoms are age-specific; that is, the affected person may have to reach his or her full height for the abnormal growth pattern to be clear, or to reach adulthood to develop some of the eye disorders associated with MFS. It is not unusual for young adult athletes to die from undiagnosed Marfan syndrome; Flo Hyman (1954–1986), who won a silver medal in volleyball in the 1984 Olympics, died suddenly of an aortic dissection during a volleyball game in 1986.

The diagnosis of Marfan syndrome is therefore based on a combination of the patient's symptoms, family history, and any information that may be obtained through genetic testing. There are several different sets of criteria that the patient's doctor may follow to arrive at the diagnosis. The four symptoms that are weighted most heavily are dilation of the aorta; dislocation of the lens of the eye; weakening of the membrane covering the spinal cord; and four or more of the skeletal changes associated with MFS.

The following tests are usually ordered as part of the diagnostic process:

- A complete physical examination that includes measuring the ratio of the patient's arm length to his or her height.
- An eye examination with a slit lamp to check for dislocation of the lens or a detached retina.
- A test for glaucoma. This test involves measuring the pressure of the fluid inside the eyeball.
- Electrocardiogram (ECG). This test measures the rhythm of the heart.
- Imaging studies of the aorta. Both computed tomography (CT) scans and magnetic resonance imaging (MRI) can be used to see whether the patient's aorta is enlarged or has begun to dissect.
- Echocardiogram. This is a test that uses ultrasound waves to produce two-dimensional images of the heart and its blood vessels.

## Treatment

Treatment of Marfan syndrome requires a team of specialists rather than just one doctor. The usual pattern is to treat health problems as they arise. A child diagnosed with MFS, for example, will usually need to see an ophthalmologist (eye doctor), pediatrician, orthodontist, and cardiologist (heart specialist). Adolescents may need to be treated by an orthopaedic surgeon if they have developed scoliosis or chest deformities. A neurologist may be consulted to treat complications involving the spinal cord.

To slow down the dilation of the aorta, even children are now given beta blockers, which are medications that cause the heart to beat more slowly and with less force. If the aorta enlarges to a certain size (about 2 inches [5 centimeters]), it is usually treated surgically. The surgeon will replace the weakened part of the aorta with a tube of synthetic material. Surgery may also be done to replace damaged or defective heart valves or to correct scoliosis or chest deformities.

People diagnosed with MFS must be careful to have regular eye examinations, echocardiograms, and other checkups intended to prevent

## WORDS TO KNOW

- **Aneurysm:** A swelling or balloon-like bulge in a blood vessel caused by weakness in the vessel's wall.
- **Aorta:** The large artery that carries blood away from the heart to the rest of the body.
- Aortic dissection: A tear in the wall of the aorta that allows blood to seep between the layers of

tissue that form the artery and push the layers apart.

- **Beta blockers:** A group of drugs given to treat abnormal heart rhythms and reduce the risk of aortic dilation in MFS patients.
- **Scoliosis:** Abnormal curvature of the spine from side to side.

the complications of the disease. They must also make a number of lifestyle adjustments, such as not smoking because of the possibility of severe lung damage. They are advised to avoid contact sports, weight lifting, and competitive sports like marathon running, although swimming, yoga, and walking are recommended forms of healthy exercise. Women with Marfan syndrome who choose to start a family must be carefully monitored throughout pregnancy because of the additional strain that pregnancy places on a woman's heart.

## Prognosis

The prognosis for MFS patients has been greatly improved by early diagnosis and treatment. Without treatment, most patients with Marfan syndrome die in their early thirties from heart problems or aortic dissection. In the early 2000s, however, the life expectancy of people with MFS has been increased to sixty to seventy years, thanks to modern heart medications and surgical techniques.

## Prevention

Marfan syndrome cannot be completely prevented because new mutations of the FBN1 gene will continue to arise. However, people with a family history of MFS should consider genetic counseling because it takes only one copy of the defective gene to produce Marfan syndrome. A parent who has Marfan syndrome has a 50 percent chance of having a child with the disorder.

### The Future

Researchers are presently studying the ways in which the FBN1 gene affects the development of connective tissue, hoping that better understanding may yield clues to new treatments. Other scientists are studying new drugs that may prevent or reduce heart complications in patients with Marfan syndrome. One specific drug that is receiving careful attention is losartan, a drug originally developed to treat high blood pressure.

SEE ALSO Cataracts; Glaucoma; Myopia; Scoliosis; Sleep apnea

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## **m** Marijuana Use

## Definition

Marijuana is a mixture of the dried leaves, stems, seeds, and buds of the hemp plant, *Cannabis sativa*. Dry marijuana is greenish or grey in color. Hashish, which contains the same mood-altering chemicals as marijuana, is a concentrated resin made from the flowering tops of hemp plants.

## Description

Marijuana is used in medicine (see sidebar) but is better known as a drug used for pleasure or to relieve stress and anxiety. It is the most widely used illegal drug worldwide. Marijuana is usually smoked in the form of a cigarette ("joint" or "reefer"), a cigar ("blunt"), or through a water pipe known as a bong. It can also be brewed as a tea or added to cookies, cakes, and other recipes. Hashish can be smoked, used as a food ingredient, or eaten alone.

The compound contained in marijuana that produces most of the plant's mood-altering effects is called THC, which is an abbreviation of its chemical name (delta-9-tetrahydrocannabinol). THC affects the parts of the brain that control memory, logical thought, muscle coordination, pleasure, the ability to concentrate, sense perception, and awareness of time. The concentration of THC in marijuana varies from 1–5 percent in most samples to as high as 15–17 percent in some specially cultivated varieties of the hemp plant. Hashish contains from 5–15 percent THC, and hashish oil contains 20 percent.

## **Demographics**

Marijuana is used as a recreational drug by people in all age groups. However, it is primarily a drug used by adolescents and young adults. Over ninety-four million Americans (40 percent) over the age of twelve have tried marijuana at least once, according to the 2003 National Survey on Drug Use and Health (NSDUH). Habitual use of the drug, however, goes up and down. In the late 1970s, more than 60 percent of high school seniors tried marijuana at least once, but this figure dropped to 33 percent by 1992. The percentage rose to 50 percent by 2002, where it has remained.

#### Also Known As

Cannabis, weed, pot, boom, Mary Jane, MJ, ganja, grass, hash, dope

#### Cause

Stress or pain relief, curiosity, peer pressure, addiction

#### Symptoms

Relaxation, drowsiness, loss of coordination, increased appetite, racing heartbeat, psychotic episodes

#### Duration

Six to twelve hours (mood); one to five days (measurable in urine)



*Marijuana plant.* SHUTTERSTOCK. About 17 percent of young adults between the ages of eighteen and twenty-five and about 4.1 percent of adults over twenty-six reported using marijuana once a month in 2005. The rates of marijuana use varied among different racial groups in the United States, with Asians reporting the lowest rate (3.1 percent) and Native Americans the highest (12.8 percent). The rates for African Americans, Caucasians, and Hispanics were 9.7 percent, 8.1 percent, and 7.1 percent respectively.

Men are more likely (10.2 percent) than women (6 percent) to use marijuana on a regular basis.

Marijuana is responsible for more than 120,000 visits to hospital emergency rooms in the United States each year. Fifteen percent of these patients are between the ages of twelve and seventeen, and two-thirds of them are male.

## **Causes and Symptoms**

The THC contained in marijuana reaches the brain via the bloodstream, either from the diges-

tive tract or from the lungs. The process is slower if the marijuana is taken by mouth than if it is smoked. It takes about half an hour for a person to feel the effects of marijuana contained in food or brewed as tea. Smoked marijuana reaches the brain in five to ten minutes. Once in the brain, THC attaches itself to nerve cells in the parts of the brain that control appetite, temperature regulation, memory, motor coordination, learning, short-term memory, sight, taste, hearing, and the ability to concentrate and think clearly.

Each intake of marijuana smoke is called a hit. If the marijuana is very strong, it may take only two or three hits for users to feel the effects of the drug. The psychological effects of marijuana last between six and twelve hours. However, the effects of the drug on motor coordination, short-term memory, and the ability to concentrate may last for as long as two days.

The effects of marijuana are not the same for all users. Depending on the strength of the marijuana, the person's emotional state at the time,

## Medical Marijuana

Marijuana has been used to treat pain and insomnia for at least 4,000 years, judging from ancient Indian and Egyptian medical texts. Marijuana use for medical purposes today, however, is controversial. It is difficult to isolate the chemicals in marijuana that relieve pain, bring sleep, and prevent vomiting from those that lead to addiction or psychotic episodes. A synthetic form of THC, the main chemical in marijuana, is used in dronabinol, a medication given to AIDS and cancer patients to relieve vomiting and increase appetite. THC helps relieve muscle cramps and spasms, as well as some symptoms of multiple sclerosis. THC can lower the fluid pressure inside the eyeball, leading some doctors to find it useful in treating glaucoma.

Several major difficulties exist in measuring the effectiveness of dried marijuana as a treatment for pain and nausea. The potency (strength) of marijuana varies considerably from sample to sample. The average sample of marijuana in the 1960s contained about 0.4 percent THC, whereas many samples in the 1990s contained between 2 and 4 percent THC. In the early 2000s, some people have produced marijuana that contains 15 or 17 percent THC. This wide variation complicates clinical studies of marijuana.

Using marijuana to treat AIDS and cancer patients is complicated because marijuana smoke



Demonstrators in San Francisco protest against federal prosecution for medical marijuana use. AP IMAGES.

irritates the lungs of some patients. Also, doctors have yet to research chemicals others than THC that are contained in marijuana. It is possible that some of these compounds could be harmful to patients already weakened by AIDS, cancer, multiple sclerosis, or other disorders.

and his or her basic physical and mental health, some users may feel relaxed and drowsy, but others feel panicky and upset. Common reactions to marijuana use include:

- Changed visual perceptions combined with the inability to judge distances accurately. This is one reason why marijuana users are at increased risk of car crashes.
- Loss of an accurate sense of time.

- Increased appetite for food.
- Impaired coordination. The National Highway Traffic Safety Administration (NHTSA) has found that marijuana has a significant effect on people's ability to drive safely for at least three hours after use. Users of marijuana are second only to alcoholics in terms of their involvement in fatal car crashes.
- Feelings of well-being, power, or importance.
- Speeded-up heartbeat.
- Dry mouth and throat.
- Dilation of the blood vessels in the eyes, giving them a reddened appearance.

About 60 percent of marijuana users have unpleasant sensations while using marijuana. Some people have strongly negative reactions to the drug, including:

- Feelings of fear, suspicion, or panic
- Feelings that the environment is unreal or that one's self is unreal
- Visual hallucinations or hearing voices
- Rapid changes in mood
- Unusual tiredness and loss of energy
- Psychotic episodes (complete loss of contact with reality)

Marijuana has a potential for addiction by itself as well as when it is used in combination with such other drugs as alcohol and cocaine. Of the estimated 6.9 million Americans who were diagnosed in 2003 as being dependent on or abusing illicit drugs, 4.2 million were dependent on or abused marijuana. Addiction to marijuana can have serious long-term consequences for a young person's success in school and the workplace.

### Diagnosis

Diagnosis of marijuana use is usually determined by a urine test for THC. In most cases, the chemical is detectable for one to five days after the person's last use of marijuana. THC does, however, remain in the fatty tissues of the body for weeks, so that some people fail drug screens for marijuana use as long as twenty-one days after their last use.

There are also blood tests that can be used to screen for marijuana use. They can also be used to distinguish between recent use of the drug and long-term excretion of THC that has been stored in the body. A more recent diagnostic technique involves analyzing a sample of the person's saliva. This test is particularly useful in evaluating people arrested for driving under the influence of marijuana.

### Treatment

About 100,000 people in the United States seek treatment each year for marijuana addiction. Treatment requires abstaining from use of alcohol, cocaine, and other drugs of abuse as well as marijuana itself. Quitting is difficult because of the side effects that many users experience when they stop using marijuana, including irritability, increased aggressiveness, anxiety, and difficulty sleeping.

They may also find it hard to stay away from peers who are continuing to use the drug or from social activities associated with using marijuana. Successful treatment usually requires a combination of medications to help the person cope with the physical effects of quitting, psychotherapy to help them deal with the emotional issues that attracted them to marijuana, and lifestyle changes to help them avoid relapsing.

### Prognosis

The prognosis for people who continue to use marijuana over long periods of time is problematic. A number of long-term risks to educational achievement, physical health, and mental health have been identified:

- Learning problems, memory problems, and falling behind in school
- · Loss of problem-solving abilities and other life skills
- Increased risk of lung infections from marijuana contaminated by fungi, and eventual lung cancer, emphysema, or other lung diseases
- Infertility
- Increased risk of psychosis
- Seizures
- Damage to the heart and cardiovascular system resulting from chronic increased heart rate and rapid changes in blood pressure
- Increased risk of accidents in the workplace or while driving caused by impaired hand-eye coordination

It is difficult to evaluate the likelihood of a person's long-term successful recovery from addiction to marijuana because many users combine

## WORDS TO KNOW

Addiction: A chronic disease characterized by compulsive drug use and by long-lasting chemical changes in the brain.	<b>Hashish:</b> A concentrated resin prepared from the flowering tops of hemp plants.
<b>Blunt:</b> A cigar that has been cut open and refilled with marijuana.	<b>Hit:</b> A single intake of marijuana smoke from a joint or bong.
Bong: A water pipe used to smoke marijuana.	<b>Joint:</b> A cigarette made with marijuana instead of tobacco.
<b>Dronabinol:</b> A medication that contains synthetic THC, given to relieve nausea	<b>Reefer:</b> Another name for a marijuana cigarette.
and improve appetite in AIDS and cancer patients.	<b>THC:</b> The abbreviation for delta-9-tetrahydrocanna- binol, the main mind-altering chemical in marijuana.

it with other drugs such as cocaine, alcohol, and amphetamines. Multiple drug use makes it difficult for researchers to determine which treatment methods have the greatest likelihood of success.

### Prevention

Educating young people about the dangers of using marijuana appears to have a limited effect in discouraging use of the drug. The best method of prevention is a healthy family life. Parents who set good examples of responsible use of alcohol and prescription drugs, and who take the time to create strong and loving relationships with their children, can lower the risk that their children will find marijuana and other mood-altering drugs attractive.

### **The Future**

At present there are no medications that specifically target marijuana addiction. Research into the effects of THC on the brain, however, is ongoing and may lead to the development of a medication that would either block the effects of marijuana on the brain or lower its appeal to people.

SEE ALSO AIDS; Alcoholism; Emphysema; Glaucoma; Smoking

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## Measles

## Definition

Measles is a highly contagious disease of the upper respiratory tract accompanied by fever and a characteristic reddish rash that spreads over the entire body. The name of the disease dates back to fourteenthcentury English and means "spot."

## Description

Measles is a disease that has been known in Europe and the Middle East for centuries. The earliest description of it comes from a Persian doctor in the tenth century named Rhazes (860–932). Measles is caused by a Also Known As Rubeola

Cause RNA virus

#### Symptoms

Fever, cough, runny nose, reddened eyes, reddish rash over entire body

## Duration

About seven to ten days

#### Measles

Measles rash on the face of a female patient. © MEDICAL-ON-LINE / ALAMY.



virus in the same group of viruses as the mumps virus. It is highly contagious and is usually spread by contact with droplets or fluids from an infected person's nose or mouth. Ninety percent of people who have not been immunized will come down with measles if they share a household with someone who has it. In countries with temperate climates, people are most likely to get measles in late winter or spring.

The disease is preceded by a prodrome or group of warning symptoms, including tiredness, fever, and loss of appetite. After an incubation period of eight to twelve days, the patient develops a higher fever of about 104°F (40°C) accompanied by an inflammation of the lining of the eyelids, sneezing, and coughing. The person can spread the disease for about two days before these symptoms appear. The characteristic rash of measles appears next, lasting for about six days. The person can continue to spread the disease for about four days after the rash appears.

Most children will feel better about ten days after the beginning of symptoms, although about one child in ten will develop an ear infection after the rash fades.

## Demographics

Measles is now a rare disease in the United States, with fewer than 100 cases in an average year. In November 2002 measles was defined as no

Measles

longer endemic (prevalent) in the United States, meaning that infected visitors to the country are the primary source of new cases. After a vaccine against measles was available in 1963, the number of new cases in the United States dropped by 99 percent by 1993. In 1997 the rate of new cases was only one case for every two million people. The last major outbreak of measles in the United States occurred from 1989 to 1991; it caused 55,000 cases and 123 deaths. In 2006 there were forty-six reported cases of measles in the United States.

In the developing world, however, measles is still a major cause of death. According to the World Health Organization (WHO), there are about thirty million new cases of measles every year, most of these being reported in Africa. Measles causes about 880,000 deaths each year, mostly in Africa and Southeast Asia. The most vulnerable patients are infants between four and twelve months of age and children infected with the HIV virus.

Measles affects all races equally. Males and females are equally likely to get measles, but girls in developing countries are more likely than boys to die from the disease. Adults who get measles are more likely than children to develop serious complications.

People at greatest risk for complications include those with weakened immune systems, including HIV infection and leukemia; those who are malnourished; those with a vitamin A deficiency; and pregnant women.

## **Causes and Symptoms**

The cause of measles is a highly contagious virus spread by droplets from the coughing

## **Measles and Native Americans**

Measles was a major cause of death among Native Americans after the early 1500s, when Europeans first came to North and Central America. Major epidemics of measles occurred in Mexico and what is now upstate New York in the 1500s; in New England in the 1600s; in New England, the Great Lakes region, the Southwest, Texas, and the Great Plains in the 1700s; and northern Mexico and the Pacific Northwest in the 1800s. In some areas nine of every ten Native Americans died during these epidemics.

Doctors have studied why Native Americans were so vulnerable to measles. Some believed it was because Native Americans had never been exposed to the disease before 1500. Another theory was that the measles virus was much stronger in the 1500s to 1800s than it is today, and that the Native Americans were overwhelmed by a more powerful infection. In the 1970s, however, Dr. Francis Black (1925– 2007), who helped develop the first measles vaccine in humans, began studying the genetic factors affecting the immune systems of the Indians in the Amazon rain forest.

Black discovered that Native Americans were more vulnerable to measles due to a relative lack of variety in their immune systems. In some parts of Africa, for example, some 200 different immune system profiles exist in the general population. So, if one person gets a viral disease, it may not spread rapidly among others because they may be resistant to the virus. Native Americans have only three different immune system profiles. This means that a disease like measles can spread much more quickly among them and cause widespread loss of life. and sneezing of an infected person. The measles virus enters the passages of the nose and upper throat until it reaches the airway and its branches into the lungs. The virus multiplies in the moist tissue lining the airway and infects the local lymph nodes, causing them to swell. It is then carried to other lymph nodes, the urinary tract, the blood vessels, the tissues lining the eyelid, and the central nervous system.

After the initial incubation period, the patient develops an itchy rash that starts on the face and head and spreads downward to cover the entire body. The rash looks like large reddish blotches that flow into each other. It eventually turns brown and looks like a stain or discoloration of the skin. The rash fades completely in about a week.

Infants and small children may also develop diarrhea and vomiting with the measles.

### Diagnosis

The diagnosis of measles is usually made on the basis of a fever lasting for three days and the appearance of reddish spots with white centers on the lining of the cheeks. These spots are called Koplik spots, named for an American pediatrician who first described them in 1896. They often appear inside the mouth about two days before the characteristic rash of measles and remain for about two days after the rash erupts.

Because measles is now rare in the United States, the doctor may take a sample of blood or nasal secretions to be tested to confirm the diagnosis.

If the doctor is concerned that the patient may have developed pneumonia as a complication of the measles, he or she may order a chest x ray. A spinal tap may be ordered if the patient has symptoms of encephalitis.

## Treatment

Treatment of measles usually consists simply of staying home to avoid spreading the disease to others, resting in bed, and drinking lots of clear fluids. Acetaminophen can be taken to bring down the fever; children should not be given aspirin, however, because of the risk of Reye syndrome. Pregnant women, infants younger than six to nine months, and people with weakened immune systems are given a shot of immune globulin, or antibodies against the measles virus. A patient who develops an ear infection or shows signs of pneumonia will be treated with an antibiotic. Young children who become severely ill with measles may be given a large dose of vitamin A.

#### Prognosis

The prognosis for most patients with measles is complete recovery after ten days to two weeks. About one person in ten will develop an ear infection, one in fifteen will develop pneumonia, and one in 1,000 will develop encephalitis (inflammation of the brain). Some patients will also develop laryngitis or bronchitis as complications of measles. Pregnant women who get measles may lose the baby, go into childbirth too early, or have a baby with an abnormally low birth weight.

The death rate from measles in otherwise healthy people in developed countries is low, about one death per 1,000 cases of the disease. In developing countries, however, the death rate may be as high as 10 percent of cases. People with weakened immune systems have a death rate approaching 30 percent.

#### Prevention

Measles can be prevented in 95 percent of people by immunization with the measles-mumps-rubella or MMR vaccine. Giving two doses of the vaccine is usually enough to prevent measles in the 5 percent who do not develop immunity with the first dose. The vaccine is usually given to children twice before they start school, the first dose at twelve to fifteen months of age and the second dose between four and six years. Infants are not usually given the vaccine because they are protected for the first six months of life if the mother was immunized.

Most people have no side effects from the MMR vaccine. About 10 percent will have a mild fever within a week of the shot, however, and about 5 percent will develop a skin rash. People who have an immune system disorder or are being treated for cancer should talk to their doctor before getting a dose of the MMR vaccine.

In recent years there have been concerns raised about a possible connection between the MMR vaccine and autism. Studies carried out by the American Academy of Pediatrics and the Institute of Medicine,

## WORDS TO KNOW

Encephalitis: Inflammation of the brain.

**Endemic:** A term applied to a disease that maintains itself in a particular area without reinforcement from outside sources of infection.

- **Koplik spots:** Small reddish spots with white centers seen on the tissues lining the cheeks in early-stage measles.
- **Prodrome:** A period before the acute phase of a disease when the patient has some characteristic warning symptoms.

however, have shown that the vaccine does not cause autism. The reason that some people thought that it might is because autism is often identified in young children at about the same time that the first dose of the MMR vaccine is usually given. Children are at much greater danger from getting the measles than from any side effect of the vaccine.

### The Future

It is possible that measles will disappear entirely in the future, as smallpox already has and as polio is likely to do. In 2001 the American Red Cross, the World Health Organization, the Centers for Disease Control and Prevention (CDC), and other public health organizations formed the Measles Initiative, whose goal was to immunize 200 million children in Africa against measles by 2006. By November 2005, the program reported that deaths from measles in Africa had already dropped by 60 percent.

**SEE ALSO** Autism; Ear infection; Encephalitis; Laryngitis; Pneumonia; Reye syndrome

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## Melanoma

See Skin cancer.



## Meningitis

## Definition

Meningitis is an infection of the meninges, the membranes that cover and protect the brain and spinal cord, and the cerebrospinal fluid (CSF) that cushions the brain. The meninges also prevent the CSF from leaking. Most cases of meningitis are caused by viruses and are usually mild; meningitis caused by bacteria, however, is a serious disease and can be fatal. Meningitis occurs when disease organisms that have entered the body and multiplied in the nose, mouth, and throat get into the bloodstream and are carried to the meninges. In a few cases meningitis can develop when the bacteria gain entrance to the body through a surgical incision or an injury to the head or neck.

Doctors sometimes divide cases of meningitis into three categories according to the speed of symptom development. Acute meningitis develops in less than twenty-four hours and is usually caused by one of several species of bacteria; it is considered a medical emergency. Subacute meningitis takes between one and seven days for symptoms to appear; it may be caused by bacteria or viruses. Chronic meningitis develops over a period of more than a week and may result from an infection or a noninfectious cause.

### Description

The central symptoms of meningitis are a high fever, headache, and stiff neck, resulting from the swelling caused by the inflammation of the meninges. Most patients also have nausea and vomiting, mental confusion, extreme sensitivity to light, chills, and rapid breathing.

Viral meningitis, which is also called aseptic meningitis, is a less severe infection than bacterial meningitis and may not require any specific treatment. It is caused by one or more enteroviruses, which are viruses that normally live in the digestive tract. Viral meningitis usually develops in the late summer and early fall, and is most likely to affect children and adults under the age of thirty. Most viral infections occur in children under the age of five. Enteroviruses are present in saliva, throat mucus, and feces; they can be transmitted through direct contact with an infected person or an infected object or surface. Viral meningitis can also be caused by the viruses that cause chickenpox, mumps, HIV infection, West Nile virus infection, and genital herpes.

Bacterial meningitis is a medical emergency and has a high mortality rate if untreated. It is spread by coughing, kissing, and contact with an infected person's saliva. There are several different types of bacteria that can cause meningitis including *Streptococcus pneumoniae*, the pneumococcus that is most common in adults, and *Neisseria meningitides*, the meningococcus that is more common in children and teenagers. Persons who have had pneumococcal meningitis may be left with lifelong damage to their nervous system that includes deafness and brain damage.

Also Known As Spinal meningitis

#### Cause

Usually viruses or bacteria

#### Symptoms

High fever, headache, stiff neck; may also include nausea, vomiting, seizures, mental confusion

#### Duration

Days to weeks; some patients suffer lifelong complications

#### Meningitis

Less common forms of bacterial meningitis are caused by *Listeria*, a bacterium that can cause the death of a baby before birth as well as meningitis; and meningitis caused by *Mycobacterium tuberculosis*, the bacterium responsible for tuberculosis. *Haemophilus influenzae*, a bacterium that was at one time thought to be the cause of flu, used to be the most common cause of bacterial meningitis in the United States; however, the introduction of a vaccine against this organism in 1990 has greatly lowered the number of cases.

Meningitis caused by fungi is rare in the general population but is a fairly common opportunistic infection in patients with AIDS.

## **Demographics**

According to the Centers for Disease Control and Prevention (CDC), there are two to three cases of bacterial meningitis for every 100,000 persons in the United States each year. About 6,000 cases of pneumococcal meningitis are reported each year, and 2,600 cases of meningococcal meningitis.

Risk factors for bacterial meningitis include:

- Age. Since the introduction of childhood vaccines, bacterial meningitis is now more common in young adults.
- Group-living situations. These may include military bases, college dormitories, and child care centers.
- Having a weakened immune system. People with AIDS or diabetes are at increased risk of meningitis.
- Working with animals. Farmers and others who work with animals have an increased risk of *Listeria* infections.
- Sex. Among newborns, boys are three times more likely than girls to get meningitis. Among adults, men and women are equally likely to be infected.



Scarring from a severe case of meningitis that nearly killed this girl at the age of eight. AP IMAGES.

## **Causes and Symptoms**

Most cases of meningitis are caused by disease organisms—viruses, bacteria, or fungi—that enter the bloodstream and are carried to the brain and the meninges. A few cases of meningitis result from inflammatory diseases like lupus or certain cancers.

The most important symptoms used to diagnose meningitis are a high fever, stiff neck, and severe headache, which may come on in less than a day after infection. Other symptoms in adults may include:

- Nausea and vomiting
- Extreme sensitivity to light
- Confusion and difficulty concentrating
- Seizures
- Loss of appetite
- Drowsiness or difficulty waking up
- Skin rash (more common with meningococcal meningitis)

Infants and small children may have somewhat different symptoms:

- Bulging of the soft spot at the top of an infant's skull
- Constant crying
- Poor feeding
- Unusual sleepiness
- Stiffness in the baby's body as well as neck

### Diagnosis

Diagnosis of the cause of meningitis is essential to proper treatment, as the antibiotics used to treat bacterial meningitis are not useful in treating viral meningitis. A patient who has acute bacterial meningitis will usually have treatment started as soon as the doctor obtains a sample of cerebrospinal fluid for testing. The CSF is obtained by performing a spinal tap, a procedure in which a needle is inserted into an area in the lower back where the doctor can easily obtain a sample of fluid.

A patient with subacute meningitis may be given a physical examination to check for an ear, throat, or sinus infection. The doctor may also perform two maneuvers to see whether the patient's meninges are inflamed. In one test, the doctor raises the patient's leg at the hip to a right angle from the examining table and tries to straighten the lower leg. If the leg cannot be straightened or the patient experiences neck pain, he or she most likely has meningitis. The other maneuver involves bending the patient's neck forward as they lie on the table. If the knees and hips flex upward, the patient probably has meningitis. The doctor may also order an x ray or computed tomography (CT) scan of the head and neck to look for swelling and inflammation.

The sample of CSF is sent to a laboratory for analysis. Identification of the specific bacterium can take several days, meanwhile, the doctor can begin to treat the patient with a broad-spectrum antibiotic until the test results come back.

In 2007 the Food and Drug Administration (FDA) approved a rapid CSF test that identifies virus particles in CSF in about three hours. This test allows doctors to distinguish fairly quickly between viral and bacterial meningitis and avoid giving unnecessary antibiotics to patients with viral meningitis.

### Treatment

It is important to get a patient with the symptoms of meningitis to a hospital as quickly as possible, particularly if the symptoms appeared in less than a day. Patients who are acutely ill and are taken to a hospital are usually treated within thirty minutes of their arrival, as emergency room doctors assume that the patient has bacterial meningitis and do not want to delay treatment until the specific organism is identified. A sample of cerebrospinal fluid is taken by a spinal tap for analysis; then the patient is given intravenous penicillin or another broad-spectrum antibiotic, intravenous fluids, and pure oxygen to assist breathing. The patient may also need to be treated for seizures, or to have fluid drained from the sinuses or from the space between the meninges and the brain. After the specific bacterium has been identified, the doctor can adjust the type and dosage of the antibiotics given to the patient.

Viral meningitis cannot be treated with antibiotics. Patients are usually advised to stay home and rest in bed for a few weeks. They can take over-the-counter pain relievers for muscle aches and pains and to bring down fever. If the viral meningitis is caused by the herpes virus, the doctor may also prescribe acyclovir, an antiviral drug used to treat herpes.

### Prognosis

Acute bacterial meningitis has a mortality rate of 10–15 percent even with treatment. Pneumococcal meningitis may have a mortality rate as

high as 21 percent. Of the patients who survive, between 10 and 20 percent will suffer such complications as blindness, hydrocephalus, hearing loss, learning disorders, or even paralysis.

Viral meningitis is usually a much milder disease. Some patients may need to be hospitalized for supportive care for a week or so, but most can recover at home within two weeks. Complications are rare with viral meningitis.

#### Prevention

There are several vaccines that can be used to prevent meningitis. As has already been mentioned, the rates of Haemophilus influenzae meningitis among young children dropped dramatically after a vaccine against this bacterium was added to childhood immunization schedules in the 1990s. There are also vaccines that have been developed to protect adults as well as children from pneumococcal and meningococcal meningitis. There is one type of pneumococcal vaccine known as PCV7, recommended for children between two and five years of age who are at high risk of infection. A different vaccine known as PPV is recommended for adults at risk of pneumococcal meningitis: those over sixty-five, those with weakened immune systems, those with diabetes or heart disease, and those whose spleen was removed. The vaccine that protects against the meningococcus is known as MCV4. It is recommended for all children at eleven and twelve and for college students who were not vaccinated at that age. MCV4 can also be used to protect people exposed to meningitis during an outbreak or who must travel to countries with high rates of meningococcal meningitis.

Other preventive measures that people can take include:

- Keep the immune system strong by getting enough sleep, exercising regularly, and eating a healthy diet.
- Wash the hands regularly, particularly when living in a dormitory or similar housing situation.
- Avoid sharing glasses, drinking cups, food utensils, and similar items with others who may be infected or exposed to infection.
- Cover the mouth or nose before sneezing or coughing.
- Take any antibiotics that may be prescribed during a meningitis outbreak in one's school or workplace.
- Ask the doctor about vaccination before traveling abroad.

## WORDS TO KNOW

Aseptic meningitis: A term that is sometimes used for meningitis that is not caused by bacteria.

**Hydrocephalus:** Abnormal accumulation of cerebrospinal fluid within the cavities inside the brain.

Meninges (singular, meninx): The membranes that cover the brain and spinal cord.

**Opportunistic infection:** An infection caused by an organism that does not cause disease in a person with a healthy immune system.

## The Future

Meningitis is likely to continue to be a health problem for the foreseeable future. There are parts of the world, such as Brazil, southeastern Asia, and Africa below the Sahara, where meningitis epidemics still occur. In addition, the bacteria and viruses that cause most cases of meningitis are commonplace organisms that are hard to avoid. The development of effective vaccines is helping to lower the rates of meningitis in the United States. At present, researchers are looking at new medications that may help to lower the rates of complications from bacterial meningitis as well as studying several new vaccines.

**SEE ALSO** AIDS; Encephalitis; Genital herpes; Hearing loss; Hydroce-phalus; Lupus

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## Mono

See Infectious mononucleosis.



## Mosquito-Borne Diseases

Mosquito-borne diseases are illnesses transmitted to humans through the bites of infected mosquitoes. The insects are called vectors of these diseases because they transmit disease-causing organisms from one host to another even though they do not cause the disease directly or become affected by the disease. For example, the mosquitoes that transmit malaria pick up the protozoa (one-celled organisms) that cause malaria from an infected human when they bite through the skin and take up blood containing the parasites. The parasites reproduce inside the salivary glands of the mosquito and are transmitted to a new host when the mosquito bites that person.

Mosquito-borne diseases include those caused by viruses as well as those caused by protozoa. There are several types of viral encephalitis (inflammation of the brain tissue), some of which can be life threatening. West Nile virus infection is another viral disease carried by infected mosquitoes.

Mosquito-borne diseases are a public health problem around the world—particularly in developing countries—because there are no effective vaccines for humans against these diseases. Prevention still depends on draining pools and water containers where mosquitoes breed; keeping mosquitoes out of the home; and avoiding traveling to countries with high rates of malaria and viral encephalitis.

SEE ALSO Encephalitis; Malaria; West Nile virus infection

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## **Motion Sickness**

## Definition

Motion sickness is a feeling of nausea or dizziness produced by conflicting information provided to the central nervous system by the various organs of sense perception. It is not a disorder or disease in the strict sense but a normal response to an abnormal situation, namely a disagreement between what the eyes see and what the organs of balance in the inner ear feel.

### Description

Motion sickness is an ancient health issue for travelers, affecting seafarers for millennia and modern travelers by automobile, airplane, or spacecraft. It can also affect people on amusement park rides or playing computer simulation games. Motion sickness can even affect people riding on an animal; for example, the famous British desert soldier, Lawrence of Arabia, often became sick when riding on camels because the swaying of the animals from side to side as they walked across the sand made him feel dizzy and nauseated.

People vary in the intensity of the symptoms they experience during motion sickness. For most people it is only a minor problem, but some people become so sick that they are almost incapacitated. One difference between the nausea and vomiting caused by motion sickness and that caused by many digestive disorders is that vomiting does not usually relieve the sick feeling in motion sickness; the person may continue to vomit without feeling better.

## Demographics

The demographics of people experiencing motion sickness vary somewhat depending on the mode of travel; almost 100 percent of people Also Known As Kinetosis, airsickness, carsickness. seasickness

#### Cause

Conflicting messages to the central nervous system from the eyes, ears, and other sensory organs

#### Symptoms

Nausea, vomiting, dizziness, cold sweats, fatigue

#### Duration

Duration of car or airplane trip; about two days for seasickness


Boy with motion sickness on a plane. MARK CLARKE / PHOTO RESEARCHERS, INC.

will get seasick on a boat in very rough waters but only about 30 percent will feel seasick sailing in relatively calm water. According to the Centers for Disease Control and Prevention (CDC), small boats and automobiles are the methods of travel most likely to produce motion sickness.

Some groups of people are more likely to develop motion sickness than others:

- Children. About 50 percent of children between the ages of two and twelve get carsick.
- People with migraine headaches.
- Women, particularly women who are pregnant or menstruating.
- Airline pilots. About 29 percent get airsick during flights; 70 percent experience motion sickness when training in flight simulators.
- Athletes. It is thought that athletes may be more susceptible to motion sickness because they have finely tuned senses and may be more aware of conflicting sensory input than most people.
- Astronauts. About 60 percent of United States astronauts experience space sickness during their first shuttle flight. Motion sickness is thought to be more common in larger spacecraft because the astronauts can move around more freely.

# **Causes and Symptoms**

The basic cause of motion sickness is a disagreement between the eyes' perception of movement and the inner ear's perception of balance and the body's orientation in space. The human sense of balance depends on the complex interaction of five different parts of the body:

• The inner ear's vestibular system. The vestibular system is a group of organs in the inner ear that provide sensory input related to movement, orientation in space, and balance. This system detects

and monitors the motions of the head, such as rotation of the head on the neck, and up-and-down, forward-and-backward, and sideto-side motions of the head.

- The eyes. The eyes help people locate themselves in space (whether they are upside down or right side up, for example) and identify the direction in which they are moving.
- Pressure receptors. These sensory receptors are located in the joints and spine; they tell the central nervous system what parts of the body are touching the ground.
- Sensory receptors in the muscles and joints. These receptors can tell what parts of the body are in motion.
- The central nervous system. The brain has to process inputs from the eyes, ears, pressure receptors, and muscle receptors, combine all this information, and interpret it.

If the combined sensory information is contradictory, the brain becomes confused. For example, someone riding in an airplane that has hit a patch of air turbulence may sense the up-and-down motion of the airplane through the pressure receptors in the body but not see any evidence of movement through the eyes. The brain receives messages that don't match or add up. One theory as to why this mismatch causes nausea and vomiting is that the part of the brain that resolves disagreements between what the eyes see and what the rest of the body feels is the same part that causes vomiting when a person eats something harmful or toxic. This area is called the area postrema and is located in the lower part of the brain stem. This theory holds that when the eyes and the inner ear send the brain conflicting messages, the area postrema decides that one of the senses is mistaken, that the mistake is due to a toxin, and that vomiting is necessary to get rid of it.

The symptoms of motion sickness include abdominal cramping, nausea, vomiting, dizziness, a pale complexion, and cold sweats. Motion sickness caused by training in a flight simulator includes eyestrain, headache, difficulties focusing or concentrating, and being unsteady on one's feet for several hours afterward. The Federal Aviation Administration (FAA) now recommends that pilots avoid flying or even driving for several hours after a simulator session.

#### Diagnosis

Diagnosis of motion sickness is usually based on the circumstances in which the person feels nauseated. Most people can tell when they are affected by motion sickness without consulting a doctor.

#### Treatment

Treatment for motion sickness usually includes preventive measures, discussed in the next section, and medications. The choice of medication depends on the length of the trip, any underlying medical conditions that the traveler may have, and concerns about drowsiness as a side effect. For example, airline pilots who have problems with airsickness are not allowed to take any medications that cause sleepiness or visual disturbances while they are in command of the plane.

Some medications for motion sickness, like Benadryl, Bonine, and Dramamine, can be taken by mouth and are available without a prescription. They should be taken between thirty minutes to an hour before the trip in order to allow them to be absorbed through the digestive tract. Scopolamine, a prescription medication, is available in both an oral form (to be taken by mouth) and a transdermal patch (the medicine is absorbed through the skin). The patch is applied behind the ear four hours before the trip and can be replaced every three days if needed. The medication in the patch that prevents nausea is absorbed through the skin. All drugs taken to prevent motion sickness, however, can cause drowsiness, dry mouth, blurred vision, and loss of coordination. People should use these medications with caution if they plan to drive, operate machinery, or go swimming or diving underwater. They should never combine these medications with alcohol.

Some types of alternative treatments that work for some people include drinking ginger tea or chewing on candied ginger. Ginger has long been used in traditional Chinese medicine to prevent nausea and is commonly recommended for morning sickness in pregnant women. Ginger also has the advantage of not causing drowsiness as a side effect. Another alternative treatment that benefits some travelers is the use of wrist bands or electric devices that stimulate a point on the wrist called the P6 point in acupuncture. The point is located about an inch and a half below the crease where the wrist meets the hand.

Most people recover from motion sickness without any difficulty. However, people who do not get better or whose symptoms get worse should see a doctor quickly—they may need specialized treatment by a neurologist (doctor who specializes in treating disorders of the nervous system) or an otorhinolaryngologist (doctor who specializes in disorders of the ears, nose, and throat). Seeing a doctor is particularly important if the patient has trouble walking, cannot see clearly, or is losing their sense of hearing.

## Prognosis

Most people who experience motion sickness feel better fairly quickly after their trip is over, although some people who suffer from severe seasickness may feel sick for two or three days after reaching land. Many people who travel frequently often develop a tolerance for the particular types of motion associated with a specific method of transportation; sailors sometimes speak of "getting one's sea legs" as a way of describing getting used to the motions of a ship without getting seasick.

#### Prevention

In addition to taking medications to prevent or minimize motion sickness, the CDC recommends the following preventive measures:

- Choose a seat that will provide a smoother ride—a seat where the eyes will see the same motion that the body and inner ears feel. These are the front seats of cars, the forward cars of trains, the center of a boat, and the seats over the wings of an airplane.
- Focus on the scenery outside the vehicle or the distant horizon rather than trying to read or looking at objects inside the vehicle.
- Minimize motions of the head and close the eyes. If possible, lie flat on the back or take a nap.
- Eat a light meal without fatty or spicy foods before traveling; do not eat a heavy meal before a trip or travel on a completely empty stomach.
- Do not drink alcoholic beverages or smoke.
- Try to minimize emotional stress and anxiety.
- If possible, open a nearby window or vent and breathe in some fresh air.

## The Future

Given the popularity as well as the necessity of travel for many people, motion sickness is likely to continue to be a common problem for

## WORDS TO KNOW

Area postrema: The part of the brain stem that controls vomiting.

- **Brain stem:** The lower part of the brain that joins the spinal cord. It controls breathing, pain perception, and other vital functions.
- **Transdermal:** Referring to a type of drug that enters the body by being absorbed through the skin.
- **Vestibular system:** The group of organs in the inner ear that provide sensory input related to movement, orientation in space, and balance.

travelers. Fortunately, it is almost always a minor inconvenience rather than a life-threatening condition.

SEE ALSO Headache

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# **U** Multiple Sclerosis

# Definition

Multiple sclerosis, or MS, is a disorder in which the nerves in the central nervous system gradually lose their protective covering of myelin, a fatty substance that insulates the nerves and allows them to transmit messages to and from the brain. The damaged myelin forms scar tissue, or sclerosis, which gives the disease its name. When the myelin is damaged, transmission of nerve impulses is not only delayed but may also be misinterpreted by the brain. The result is an assortment of visual disturbances, problems with movement, hearing difficulties, fatigue, strange sensations in the arms and legs, and other symptoms that vary from patient to patient. MS is an autoimmune disorder, which means that it is caused by the body attacking itself.

# Description

One of the most confusing aspects of MS is that some patients are only mildly affected by the disease for many years, while others become almost completely disabled over time. To help explain the different forms that multiple sclerosis may take, the National Multiple Sclerosis Society standardized four subtypes of MS in 1996. These subtypes are used to determine treatment and predict the future course of the disease:

- Relapsing-remitting (RR). About 85 percent of patients with MS have this form of the disease. The patient has occasional attacks (also called relapses) in which symptoms get worse, followed by periods of remission in which the patient has stable health.
- Primary progressive (PP). About 10 percent of MS patients are in this category. In PP multiple sclerosis, the disease gets worse over time from the beginning with no distinct relapses or remissions.
- Secondary progressive (SP). Patients with the SP form of the disease start out with the relapsing-remitting form and then develop the primary-progressive form. Between half and three-quarters of

Also Known As MS, disseminated sclerosis

#### Cause

Unknown; possibly a combination of genetic and environmental factors

#### Symptoms

Difficulties with vision, hearing, coordination, thinking; numbness or tingling; fatigue

Duration Years



patients with RR multiple sclerosis develop the SP form within ten years. It is not yet known whether recently introduced drugs to treat MS will change this pattern. SP is the subtype that causes the greatest disability.

• Progressive relapsing (PR). About 5 percent of patients with MS have the progressive relapsing type. They get steadily worse from the beginning with periodic attacks of more severe symptoms along the way.

In addition to these four subtypes of MS, there is also a category called clinically isolated syndrome or CIS. It is used to refer to people who most likely have relapsing-remitting MS but have had only one episode of illness and do not yet meet all the criteria for a diagnosis of MS. People with a CIS may or may not go on to develop MS.

Image of the brain of a patient with damage from multiple sclerosis. © PHOTOTAKE INC. / ALAMY.

# **Demographics**

According to the National Institute of Neurological Disorders and Stroke (NINDS), there are between 250,000 and 300,000 people who have been diagnosed with MS in the United States, with about 200 cases diagnosed each week. There are likely to be many others, however, in the early stages of the disease whose symptoms are too mild or too uncertain to lead to a diagnosis of MS. Worldwide, the rate of MS varies from country to country, ranging between two and 150 per 100,000 people; it is estimated that there are 2.5 million people around the world with MS.

Multiple sclerosis is almost entirely a disease of adults. It is most likely to affect people between the ages of twenty and fifty, with the average age at onset being thirty-four years. It is more common among Caucasians of northern European background than among African Americans or Native Americans. Some ethnic groups, such as the Inuit of Alaska and the Maoris of New Zealand, rarely develop MS. MS is twice as likely to affect women as men. Men, however, are more likely to develop the primary progressive form of the disease.

# **Causes and Symptoms**

Although researchers know that the symptoms of MS are caused by damage to the myelin coverings of nerve fibers, they have not been able to determine what causes the damage to the myelin initially. One theory holds that infection by some kind of virus turns the body's immune system against the myelin sheaths along the nerve endings. Other researchers think that it may be a bacterial infection that triggers MS. Still another theory involves a possible role of vitamin D in the development of the disease. Vitamin D is produced in the body by exposure to sunlight, which is more abundant in countries closer to the equator than in those closer to the poles. People in the tropics have higher levels of vitamin D in their bodies and lower rates of MS. It is thought that this vitamin boosts the immune function and may help protect against autoimmune diseases like MS.

In addition to possible infectious and environmental causes, researchers think that genetic factors play some role in MS even though the disease is not directly inherited. Although the risk of developing MS in the general population is one in 750, the risk rises to one in forty in anyone who has a first-degree relative (parent, sibling, or child) with the disease. An identical twin of a patient with MS has about one in four chance of developing the disease.

The symptoms of MS may include some or most of the following, depending on which parts of the nervous system have been damaged by the loss of myelin:

# A Writer Reflects on Life with MS

Joan Didion (1934–) is an American writer of novels and screenplays who was diagnosed with MS in the 1980s. Although she has a relatively mild form of the relapsing-remitting form of the disease, she observes that the emotional impact of MS can be intense: "The knowledge that one has a potentially serious and debilitating disease—no matter how benign a form it seems to be taking—does change one's life and requires a significant adjustment.... One can certainly overreact or make the disease the centerpiece of one's life, and I am by no means advocating that, but the difficulty of coming to terms with it should not be understated."

Didion continues: "The primary task [for me] was learning to live with day to day uncertainty. Uncertainty about tomorrow requires more immediate adjustment. It is easy to feel anger and frustration when, because one's physical capacity varies so from day to day, plans cannot always be carried out. It becomes even more necessary than usual to know one's priorities. The need to establish priorities is common to everyone but becomes more acute under the pressure of this level of daily uncertainty. I also discovered that my expectations of what I should be capable of were getting in my way. I had to learn to adjust those expectations to the reality of what I was capable of and, more important, to understand that who I was did not depend on what I could do physically."

- Fatigue. It is not unusual for patients with MS to be too weak to carry groceries upstairs or go for a walk. Fatigue affects about 80 percent of patients with MS.
- Visual problems, including seeing double, blurry vision, or pain in the eyes. Visual problems are often the first noticeable symptom of MS.

- Problems with gait, balance, and coordination.
- Bladder and bowel dysfunction, including constipation, diarrhea, and inability to control urination.
- Dizziness.
- Changes in mental function, including memory loss and difficulty concentrating.
- Sexual dysfunction and loss of interest in sex.
- Depression and rapid mood changes.
- Spasticity. Spasticity refers to muscle stiffness and involuntary spasms of the muscles.
- Numbness in the limbs or face. Some patients may also experience tingling or itching sensations.
- Less common symptoms of MS include loss of hearing; headaches; speech disorders; swallowing problems; and seizures.

## Diagnosis

There is no single test that can be used to diagnose MS. It is essentially a diagnosis of exclusion, which means that the doctor arrives at the diagnosis by first ruling out other possible causes of the patient's symptoms. The doctor usually begins by taking a complete history of the patient's symptoms and looking for a possible pattern in their occurrence. Other tests that may be used to clarify the diagnosis include:

- A neurological examination. The doctor will test the patient's gait (habitual manner of walking), reflexes, muscle tone and strength, perceptions of heat, cold, and vibration, and coordination and balance.
- Psychiatric examination. This may be given to test for memory loss and other disorders of thought. It may also be given to rule out depression or other psychiatric disorders that may cause fatigue, loss of sexual interest, and other symptoms experienced by some patients with MS.
- Multiple resonance imaging (MRI). This imaging technique can reveal lesions in the brain caused by the loss of myelin.
- Spinal tap. This is a procedure in which the doctor withdraws a sample of cerebrospinal fluid through a needle inserted into the spinal column. It can be used to look for an abnormally high level of white blood cells and certain proteins that are characteristic of MS, or to rule out other possible disorders of the central nervous system.

• Evoked potential (EP) testing. These are tests that record the nervous system's electrical responses to specific forms of sensory stimulation, usually visual, hearing, and general sensory perception.

After ruling out other possible causes of the patient's symptoms, the doctor can base a diagnosis of MS on showing that damage has occurred to the myelin in at least two separate areas of the central nervous system and that the areas of damage occurred at least one month apart.

# Treatment

There is no cure for MS. The disease is managed by the use of drugs intended to modify the course of the disease; treat initial symptoms and relapses; improve the patient's safety and ability to function through physical rehabilitation; and provide emotional support.

Therapy for MS may include:

- Disease-modifying drugs. There are six of these that were approved by the Food and Drug Administration (FDA) as of 2008. These compounds are intended to slow the progression of MS and postpone relapses. They cannot, however, be taken by women who are pregnant or planning a pregnancy.
- Drugs to treat specific symptoms. Spasticity can be treated with muscle relaxants; visual symptoms often respond well to a drug called Solu-Medrol; fatigue can be treated by antidepressants; and muscle or back pain can be treated with aspirin with or without codeine.
- Relapses are most often treated with cortisone and other steroid medications.
- Rehabilitation for patients with MS may include several types of therapy depending on the patient's specific symptoms. Occupational therapy, physical therapy, speech therapy, and training for a different occupation may all be needed.

Patients with MS are often advised to avoid hot showers, saunas, or hot tubs, and vigorous exercise, as heat appears to trigger relapses of the disease. Those who live in hot, humid climates should have an air conditioner in at least one room of their house.

Some patients with MS benefit from complementary and alternative (CAM) treatments, including yoga, relaxation techniques, hypnosis, meditation practice, pet therapy, and humor therapy.

# WORDS TO KNOW

**Clinically isolated syndrome (CIS):** A term applied to patients who have had one episode of illness that suggests they have MS but do not yet meet the full criteria for diagnosis.

**Myelin:** A fatty substance that insulates nerve fibers and allows for speedy and accurate transmission of nerve impulses.

**Progressive:** A term that refers to a disease that gets worse over time. MS is a progressive disease.

**Relapse:** A return or recurrence of the symptoms of a disease.

**Remission:** A period of relief from the symptoms of a disease.

Sclerosis: Hardening or scarring of tissue.

Spasticity: Stiffness or spasms in the muscles.

#### Prognosis

The prognosis for MS varies; most patients are relatively symptom-free and able to complete school or work for almost twenty years. The patients with a more favorable prognosis are those who are female; those who are younger than thirty when their symptoms started; those who have infrequent attacks; those who have the relapsing-remitting form of the disease; and those who have less severe symptoms.

Patients are more likely to die from complications associated with MS than from the disease itself. However, it is estimated that the disease shortens a patient's life span by about seven years.

#### Prevention

There is no known way to prevent MS.

## The Future

Researchers are working on a definitive diagnostic test for MS as well as better treatments and an improved understanding of the role of the immune system in the disease process. Some scientists think that identification of the genetic factors involved in the disease combined with further study of environmental factors may help find ways to prevent the disease.

**SEE ALSO** Cerebral palsy

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# Muscular Dystrophy

# Definition

Muscular dystrophy, or MD, is actually the name of nine groups of inherited disorders that affect muscle strength and the connections between

#### Muscular Dystrophy

In this patient with muscular dystrophy, weakness in the shoulder muscles causes the shoulder bones to stick out. © MEDICAL-ON-LINE / ALAMY.



muscles and nerves. Some begin in childhood while others affect mostly adults; some lead to early death while others cause only mild disabilities. What they have in common is progressive weakening of the skeletal muscles. Some forms of MD also affect the muscular tissue of the heart.

## Description

The nine types of MD vary in the age of onset, major symptoms, and course.

- Duchenne muscular dystrophy. Named for Guillaume Duchenne (1806–1875), the French doctor who first described it in the 1860s, Duchenne muscular dystrophy is the most common form, accounting for half of all cases of MD. It is gender-linked, affecting mostly boys, and is usually diagnosed in childhood. Muscle wasting begins in the muscles of the legs and pelvis, then moves to the arms and neck. Boys with Duchenne MD have frequent falls, breathing difficulties and heart problems, and are usually confined to a wheel-chair by age twelve. Most patients die before age twenty.
- Becker muscular dystrophy. This is a milder form of MD but affects the same muscles as Duchenne muscular dystrophy. Patients are generally diagnosed at age eleven but generally remain independent longer than patients with Duchenne MD and live into middle age.

Also Known As

MD, inherited myopathy

#### Cause

Mutations in various genes that affect the production of muscle proteins

#### Symptoms

Loss of muscle strength and tissue, falls, scoliosis, joint contractures, heart problems

#### Duration

Lifelong

- Congenital MD. This is a group of muscular dystrophies that affects both boys and girls and is usually diagnosed before age two. Some children with these disorders die in infancy while others live into adulthood.
- Emery-Dreifuss MD. This form of MD primarily affects boys; symptoms appear at any point from age ten to twenty-five. Contractures in the ankles, knees, and other joints occur before the muscles start to weaken. Almost all patients develop heart problems by age thirty.
- Facioscapulohumeral MD. This form of muscular dystrophy affects the muscles of the face, shoulders, and upper arms before other parts of the body. Symptoms may begin at any age from the late teens through age forty. Patients often develop a pouting or masklike expression as the muscles of the face deteriorate.
- Limb-girdle MD. This form of MD affects both boys and girls. Muscle weakness is typically noticed first around the hips before spreading to the shoulders, legs, and neck. People with this form of MD develop a waddling gait, fall frequently, and are unable to run.
- Distal MD. This form of the disease does not appear until patients are forty to sixty years old. It affects the muscles of the hands, fore-arms, lower legs, and feet.
- Myotonic MD. This is the most common form of adult-onset MD, appearing between twenty and thirty years of age. It affects both men and women. Also known as Steinert's disease, this form of MD is characterized by stiffness; the patient cannot relax the muscles voluntarily after tightening then. In addition to weakening the patient's muscles, myotonic MD affects the central nervous system and other organs, including the heart, adrenal glands and thyroid, the eyes, and the digestive tract. People with myotonic MD may also develop cataracts in the eyes.
- Oculopharyngeal MD. This form of muscular dystrophy typically begins when the patient is in his or her forties or fifties. It also affects both men and women. It is most common in people of French Canadian descent and some Hispanics living in New Mexico. Muscle weakness first affects the eyelids and throat muscles, causing difficulty swallowing as well as interfering with vision. Most patients will eventually lose the ability to walk.

#### Demographics

The demographics of muscular dystrophy are different for different forms of the disease. Duchenne MD is the most common, occurring in about one in every 3,500 male babies. Becker MD is the second most common, with a rate of one in every 30,000 male births. The other forms of MD are very rare; limb-girdle dystrophy, for example, occurs in only 1 percent of patients diagnosed with muscular dystrophy, while Emery-Dreifuss MD affects only one boy in every 100,000. Oculopharyngeal MD is most common among French Canadians and some Hispanics, while distal MD is more common in Sweden than in other countries.

As has been noted, some forms of MD affect only males because the genetic mutation responsible for the absence or defective quality of a protein essential to maintaining muscle tissue is located on the X chromosome. Females may be carriers of the mutation and may have mild symptoms of the disorder. Other types of muscular dystrophy have different inheritance patterns and can affect women as well as men.

#### **Causes and Symptoms**

The basic cause of the various forms of MD is a genetic mutation that affects the production of proteins necessary to maintain the structure of muscle cells. In the case of Duchenne MD and Becker MD, the affected gene is called the DMD gene and is located on the X chromosome. The gene is responsible for the production of a protein called dystrophin, which is needed to maintain the cell membrane of muscle cells. In Duchenne MD, there is no dystrophin produced; in Becker MD, there is some dystrophin, but not enough to protect the muscle fibers from gradually weakening.

In Emery-Dreifuss MD, the defective gene on the X chromosome is called the EMD gene. It is responsible for producing a protein called emerin, which is essential for the functioning of skeletal and heart muscles. Myotonic dystrophy is caused by a defect in a gene on chromosome 19. The defect interferes with the production of a protein called myosin, which is involved in the movement of muscles.

Although there are some variations among the nine types of muscular dystrophy, the most common symptoms of the disease include:

- Muscle weakness that gets worse over time
- Contractures
- Loss of balance and coordination

- Wasting (loss of muscle tissue, replaced by fat in some cases)
- Scoliosis (curvature of the spine)
- Difficulty walking
- Frequent falls
- Clumsiness; eventual loss of the ability to write
- Gradual weakening of the heart and breathing muscles
- Eventual difficulty in swallowing

# Diagnosis

Since the symptoms of MD appear in different patients at different ages, depending on the specific type of disease that the patient has, diagnosis begins with a physical examination and taking a history of the patient's symptoms. Next, the patient may be sent to a neurologist (a specialist in diagnosing and treating disorders of the nervous system) for tests of the reflexes, balance, and other functions in order to rule out exposures to toxins or other diseases that may be causing the patient's symptoms.

The third step is a series of laboratory and imaging tests:

- Blood and urine tests. These are used to measure the levels of various enzymes in the patient's body fluids that result from the breakdown of muscle tissue or that leak out of damaged muscle. A blood test can also be performed for genetic testing.
- Exercise tests. These can be carried out at the patient's bedside. They measure the strength of the muscles, particularly the muscles involved in breathing.
- Ultrasound imaging. Ultrasound imaging can detect abnormalities in the patient's muscles even in early stages of MD. Magnetic resonance imaging (MRI) can also be used to look for changes in muscle tissue and to monitor the progression of the disease.
- Muscle biopsy. Taking a sample of the patient's muscle tissue allows the doctor to distinguish between MD and other diseases that affect the muscles and nerves.
- Electromyography. Electromyography is a technique in which the doctor inserts a thin electrode through the skin into a muscle. As the patient tightens and relaxes the muscle, the electrode measures the patterns of electrical activity in the muscle.

- Electrocardiogram (ECG). This test is done to evaluate the functioning of the patient's heart.
- Pulmonary function test. The doctor may order this test for patients whose breathing muscles are showing signs of weakness. It measures the patient's lung capacity.

# Treatment

There is no cure for muscular dystrophy. Treatment consists of keeping the patient mobile and comfortable as long as possible. Specific therapies may include medications to delay the loss of muscle tissue and function or to relax tightened muscles; surgery to minimize problems caused by contractures or scoliosis; physical therapy to keep the muscles as strong and flexible as possible; and occupational therapy to teach the patient the skills that are necessary for self-care. Some patients with breathing difficulties may be placed on an artificial respirator, while those with heart problems may have a pacemaker implanted.

Most patients with MD eventually require assistive devices, from canes, braces, and walkers to wheelchairs and motorized scooters. These devices can help to keep patients independent as well as slow the development of contractures. It is important for patients with MD to stay as active as possible, as bed rest or inactivity can speed up the deterioration of the muscles.

# Prognosis

The prognosis of muscular dystrophy depends on the specific type, although all types are incurable. Patients with Duchenne MD rarely live beyond age twenty, while those with congenital MD may die in childhood, and patients with one of the adult-onset dystrophies may live into their sixties. Modern medical and surgical care allows most patients a better quality of life than they would otherwise have, but the outcome is still premature death, usually from heart failure.

# Prevention

There is no known way to prevent all the various types of MD. Genetic counseling is advised, however, when there is a family history of muscular dystrophy. Women may have no symptoms but still carry the gene for the disorder, and Duchenne muscular dystrophy can be detected by genetic tests during pregnancy.

# WORDS TO KNOW

**Contracture:** Shortening or tightening of the muscles surrounding certain joints that limits the movement of the joints.

- **Dystrophin:** A protein found in muscle whose absence or defectiveness is one of the causes of muscular dystrophy.
- **Gene therapy:** The insertion of normal genes into a person's cells or tissues in order to treat a disease by replacing a harmful mutation or nonfunctioning gene.

Myosin: A protein involved in muscle movement.

**Nucleotides:** The basic structural units of DNA and RNA.

Scoliosis: Curvature of the spine from side to side.

**Triplet:** In genetics, a unit of three nucleotides that starts or stops the production of a specific protein. Triplets are also called codons.

Wasting: Loss of lean muscle tissue.

# The Future

As of 2008, a drug known as MYO-029 was being tested in clinical trials as a possible treatment for Duchenne MD. Researchers think that this drug might help damaged muscle to repair itself, and thus slow down or even stop the gradual loss of muscle tissue. Another approach that has been tried in treating MD is gene therapy, in which the defective DMD gene could be replaced by a normal gene. New strategies for replacing the defective gene are the target of current research.

Research into the causes of and possible treatments for MD received a major boost in December 2001, when President George W. Bush signed into law the Muscular Dystrophy Community Assistance, Research, and Education (MD CARE) Amendments Act of 2001. The act led to the establishment of six muscular dystrophy research centers at university medical centers and children's hospitals across the United States.

**SEE ALSO** Cataracts; Scoliosis

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# Myopia

# Definition

Myopia is a refractive error (inability to focus clearly) in which people can see near objects clearly but not distant ones, which usually look blurry. It is not an eye disease in the strict sense.

# Description

Myopia or nearsightedness is a common refractive error in which a person cannot see distant objects clearly. In most cases the problem is caused by an abnormally long eyeball, a steeply curved cornea, an overly thick lens, or a combination of all three factors. The length of the eyeball or curvature

#### Also Known As Nearsightedness, shortsightedness

#### Cause

Long eyeball, thick lens, steep cornea, or a combination of all three

#### Symptoms

Inability to see distant objects clearly

#### Duration

Lifelong unless corrected by surgery

#### Myopia



*Vision in a normal eye (top) and in a myopic eye.* THE GALE GROUP.

of the cornea or lens causes images to be focused in front of the retina (the light-sensitive tissue at the back of the eye) rather than on it.

People with myopia not only need prescription lenses for nearsightedness when they are young but may also require bifocals as they age because the eye gradually loses its ability to accommodate (change focus) as a person gets older.

#### Demographics

Myopia is a common refractive error worldwide, although the rates vary from country to country and from age group to age group. According to a British medical journal, the prevalence of myopia (including mild cases that do not require the person to wear glasses) has been reported as high as 70–90 percent in East Asia and Japan, 30–40 percent in Europe and the United States, and 10–20 percent in Africa. It is estimated that 44 percent of Japanese and Taiwanese adults have myopia severe enough to require corrective lenses, whereas in India, the rate is 7 percent of the general adult population. A study of British university students showed that 50 percent.

In the United States, the rate of myopia in the general population severe enough to require corrective lenses is thought to be between 20 and 25 percent. Although a few children are born with myopia, the condition is most likely to appear between ages five and twenty. About 25 percent of Americans in this age group are nearsighted. About 26 percent of people in the United States between the ages of twelve and fifty-four have myopia severe enough to require correction.

Ethnicity makes a difference in rates of nearsightedness in the United States. About 78 percent of Asian Americans have myopia, followed by Hispanics (13 percent), African Americans (7 percent), and Caucasians (5 percent). As far as is known, myopia is equally common in men and women.

# Seven Centuries of Eyeglasses

Many people think of eyeglasses (or spectacles) as a modern invention when in fact they date back to the late thirteenth century. Salvino d'Armate (1258– 1312), an Italian living in Florence, is credited with inventing the first spectacles around 1284. These early glasses did not have temple bars like presentday models but perched on the bridge of the wearer's nose. The first portrait of a person wearing eyeglasses—a clergyman reading in his library—was painted in France in 1352. The earliest eyeglasses were intended to correct farsightedness. It was not until the fifteenth century that eyeglasses were created for people with myopia.

Eyeglass development increased rapidly after 1604, when Johannes Kepler (1571–1630), a German mathematician and astronomer, published the first explanation of how properly shaped glass lenses can correct farsightedness and nearsightedness. American statesman Benjamin Franklin (1706–1790), who suffered from both farsightedness and nearsightedness in midlife, invented bifocals in 1784 when he grew tired of having to switch between two pairs of eyeglasses. British astronomer George Airy (1801– 1892) created the first lenses to correct astigmatism in 1825. The first contact lenses were developed in Germany in 1892, although these early models were large by modern standards and could not be worn comfortably for more than several hours at a time.



Johannes Kepler.

Persons with myopia in the twenty-first century have a wide variety of eyeglasses, contact lenses, and surgical treatments available for vision correction. More flexible frames, designer styling, and the use of lighter shatterproof plastic lenses instead of glass make eyeglasses increasingly attractive and comfortable to wear.

#### **Causes and Symptoms**

Myopia is thought to result from a combination of genetic factors and close visual work over an extended period of time in childhood. Myopia is known to run in families. In addition, the different rates of myopia among different races and ethnic groups points to some kind of genetic cause. More recently, the PAX6 gene on chromosome 11 has been identified as a gene that appears to affect the length of the human eyeball.

Some doctors refer to the development of myopia in the elementary grades as school myopia, as the close work involved in learning to read and write appears to trigger nearsightedness in about 10 percent of children in the United States.

The symptoms of myopia often emerge during a child's first years in school. Parents may notice that the child holds a book very close while reading or leans close to the desk surface while writing. He or she may squint a lot or sit very close to the television or blackboard. Other symptoms include headaches and failure to notice distant objects.

#### Diagnosis

Myopia and other refractive errors are evaluated by a series of vision tests. After the examiner takes a history of the patient's symptoms (including a family history of eye problems), the patient is usually asked to read the letters on an eye chart known as a Snellen chart. Each eye is tested separately. The examiner may also shine lights into the eyes or administer eye drops that allow him or her to see all the structures inside the eye clearly. This part of the examination allows the doctor to evaluate the severity of the patient's nearsightedness.

To measure the strength of the lens needed to correct the patient's myopia, the examiner uses a device called a photopter (or refractor). The photopter is placed in front of the patient's eyes and the examiner moves various lenses in and out of the device while the patient rereads the letters on the Snellen chart. The photopter can also be used to measure the correction needed for a bifocal lens.

## Treatment

Very mild myopia may not need corrective treatment. A person should see an eye doctor, however, if he or she is developing headaches or eye strain, or if blurry vision is interfering with daily activities.

People whose nearsightedness is severe enough to require correction have several options:

- Eyeglasses. These can be used to correct nearsightedness caused by uneven curvature of the lens or cornea as well as the length of the eyeball. Eyeglasses are prescribed by an optometrist or ophthalmologist but made and fitted by an optician.
- Hard contact lenses. These usually provide more effective correction of nearsightedness than soft contact lenses.
- Orthokeratology (Ortho-K). This is a procedure in which the person wears hard contact lenses for several hours overnight in

order to gradually correct the curvature of the cornea. The lenses are removed during the day. Ortho-K is also referred to as corneal molding. It does not permanently improve vision. If the patient stops wearing the retainer lenses, his or her vision may return to its original condition. Ortho-K, however, is ineffective in correcting myopia caused by an abnormally long eyeball.

- Laser surgery. If a person's nearsightedness is related to the shape of the cornea, an ophthalmologist can use lasers to reshape the cornea either by making a flap in the surface of the cornea and reshaping the tissue of the cornea under the flap, or by completely removing the upper layer of tissue in the cornea before reshaping the lower layers of tissue.
- Lens implantation. Lens implantation is a controversial treatment for moderate or severe myopia. The ophthalmologist surgically inserts a clear corrective lens inside the eye in front of the natural lens. The procedure was not performed very frequently in the early 2000s, however, because it has a high risk of complications.

There are drawbacks to surgical correction of refractive errors. These include the risks of infection, development of haze in the cornea, or dry eyes. In some cases the surgeon may need to perform a second operation if the first one either overcorrected or undercorrected the shape of the patient's cornea.

It is important for a patient with myopia to discuss all the treatment options with the optometrist or ophthalmologist, as no two people have exactly the same degree of visual blurring or the same lifestyle. In addition, patients with diabetes require very careful evaluation before any type of eye surgery because diabetes weakens the retina of the eye and increases the risk of glaucoma.

## Prognosis

The prognosis of myopia depends partly on its severity. People with any degree of myopia can have their vision corrected satisfactorily by eyeglasses, contact lenses, or surgery. People with severe myopia (about 30 percent of nearsighted patients), however, have an increased risk of retinal disorders and glaucoma after age forty. They should therefore schedule regular eye examinations to reduce the risk of these complications.

#### Prevention

Myopia is still largely considered a matter of heredity and cannot be prevented by any method known. There have been various attempts to slow

# WORDS TO KNOW

- Accommodation: The medical term for the eye's ability to change its focus automatically for viewing objects at different distances.
- **Astigmatism:** A refractive error caused by irregularities in the shape of the cornea or the lens of the eye.
- **Cornea:** The transparent front part of the eye where light enters the eye.
- **Ophthalmologist:** A doctor who specializes in diagnosing and treating eye disorders and can perform eye surgery.
- **Optician:** An eye care professional who fills prescriptions for eyeglasses and corrective lenses.
- **Optometrist:** An eye care professional who diagnoses refractive errors and other eye problems and prescribes corrective lenses.

- **Photopter:** A device positioned in front of a patient's eyes during an eye examination that allows the examiner to place various lenses in front of the eyes to determine the strength of corrective lenses required.
- **Refractive error:** A general term for vision problems caused by the eye's inability to focus light correctly.
- **Retina:** The light-sensitive layer of tissue at the back of the eyeball.
- **Snellen chart:** A series of letters arranged in lines on a chart to be viewed from a distance of 20 feet (6 meters). It is used to measure how well someone can see.

the progression of nearsightedness in schoolchildren by eye exercises or such alternative therapies as biofeedback, but none have proved to be successful. People can, however, live comfortably with nearsightedness by visual screening in childhood, regular eye checkups at all ages, and wearing corrective lenses when needed.

# The Future

Nearsightedness is such a common refractive error in the general population that it is not likely to disappear in the near future. It is possible that the present variety of treatment options will be improved or expanded by further research.

SEE ALSO Astigmatism; Glaucoma; Hyperopia

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# Ν



# Genetic

Infection

Multiple

Other

Injury

Unknown

#### Also Known As

Excessive daytime sleepiness

#### Cause

Inability of the brain to regulate the sleep/wake cycle

#### Symptoms

Daytime sleepiness, cataplexy, sleep paralysis, hallucinations when awaking

#### Duration

Years



## Definition

Narcolepsy is a sleep disorder caused by the brain's inability to regulate normal sleep/wake cycles. People with narcolepsy have sleep episodes in the daytime, hallucinations when falling asleep or awaking, and often awaken at night.

# Description

Narcolepsy can affect people in any age group but may not be diagnosed for as long as ten years after onset. It can be a disabling condition. Because people with narcolepsy can fall asleep in the daytime in the middle of other activities—including eating, driving, or operating heavy equipment—they can be involved in serious accidents. Children or adolescents with narcolepsy can fall behind in school or be judged as lazy students even though they are of normal intelligence.

Having narcolepsy does not mean that people with the disorder sleep more than other people. The disorder is better understood as a disturbance of the normal boundaries between sleeping and waking rather than simply sleeping too much. A normal adult who sleeps for about eight hours has between four and six sleep cycles during that time. A sleep cycle is a period of a type of sleep called non-rapid eye movement (NREM) sleep followed by a period of rapid eye movement (REM) sleep. REM sleep is

#### Narcolepsy

People with narcolepsy can face harm if they fall asleep in a dangerous situation, such as driving a car. PHOTOGRAPHY BY LEITHA ETHERIDGE-SIMS.



characterized by increased brain activity, rapid movement of the eyes below the lids, and sleep paralysis. People who say that they were having a dream when they woke up have been awakened from REM sleep. In normal adults, a sleep cycle is about 100–110 minutes long, 80–100 minutes of NREM sleep followed by 10–20 minutes of REM sleep.

In narcolepsy, however, the patient's sleep cycles do not follow this normal pattern. Instead, the person falls into REM sleep very shortly after falling asleep. In addition, the short periods of sleep that the patient has during the daytime are REM sleep. This abnormal occurrence of REM sleep helps to explain such symptoms of narcolepsy as sleep paralysis and cataplexy.

## **Demographics**

It is possible that narcolepsy affects more people than is presently known. The rate of narcolepsy in the general population varies widely around the world. In Israel, for example, only about one person per 500,000 is diagnosed with narcolepsy whereas the rate in Japan is one person in every 600. In the United States, narcolepsy affects about one person in every 2,000. It is slightly more common in African Americans than in members of other races.

Narcolepsy affects men and women equally. It is not thought to be inherited. However, about 10 percent of Americans with narcolepsy have a relative with the condition, but this percentage is low compared to other disorders that are known to be purely genetic in origin.

Most cases of narcolepsy first appear in people between the ages of ten and twenty-five years, but the disorder has been diagnosed in children as young as three and in adults over fifty.

#### **Causes and Symptoms**

The basic cause of narcolepsy remains unknown. Progress has been made since the late 1990s in identifying two pieces of the puzzle. The first is a connection between narcolepsy and changes in a set of genes on chromosome 6 called the HLA complex. It is thought that changes in these genes may be linked to the loss of cells in the brain that secrete hypocretin, a protein that regulates the sleep/wake cycle. Researchers do not know exactly what causes the loss of the brain cells that make hypocretin. Because the HLA complex has been linked to other autoimmune disorders, however, some doctors think that narcolepsy may be an autoimmune disease. No one has been able to prove this theory as of late 2008, however. Other researchers think that the hypocretin-producing cells may be damaged by some type of infection, but this theory has yet to be proved.

Narcolepsy has four primary symptoms:

- Excessive daytime sleepiness (EDS). This symptom may take the form of "microsleeps" of a few seconds, a long-lasting feeling of drowsiness, or daytime episodes of sleep lasting half an hour or longer.
- Cataplexy. Cataplexy refers to a sudden loss of muscle tone that may result in slurred speech, sagging of the facial muscles, general muscle weakness, inability to hold up one's head, buckling of the knees, or loss of strength in the arms. Cataplexy is often triggered by strong emotions, whether positive feelings like laughter or pleasant surprise or negative emotions like fear, shock, or anger. Cataplexy can last from a few seconds to as long as thirty minutes. The person is awake and alert during an episode of cataplexy even though he or she may appear unconscious to others.
- Hypnagogic hallucinations. Hypnagogic refers to the period of partial alertness that people have when they are waking up or

falling asleep. People with narcolepsy experience very vivid and sometimes frightening dreams when they are half awake, and they may mistake these dreams for reality.

• Sleep paralysis. Sleep paralysis refers to a temporary inability to move or speak while falling asleep or waking up. These episodes last only one or two minutes and do not affect everyone with narcolepsy, but they can be frightening to patients who do have them.

Other symptoms experienced by some people with narcolepsy include automatic behavior, in which the person performs a routine task like sorting laundry or making a cup of coffee without conscious awareness or later memory of the action. People with narcolepsy may also thrash about in bed at night or act out their dreams by screaming or waving their arms.

#### Diagnosis

The diagnosis of narcolepsy is often delayed because its symptoms resemble those of such other disorders as depression, seizure disorders, simple lack of sleep, or even illegal drug use. Early diagnosis is important because the impact of the disorder on a person's education, employment, relationships, and self-esteem can be severe. According to one study, 24 percent of adults with narcolepsy had to quit working and 18 percent had been fired from their jobs because of the disease.

The diagnosis of narcolepsy is based on a combination of the patient's history, the results of a screening questionnaire, and overnight testing in a sleep laboratory. The screening questionnaire that is used most commonly is the Epworth Sleepiness Scale, or ESS, which was developed in Australia in the early 1990s. The patient may also be asked to keep a sleep diary for one or two weeks and wear a device called an actigraph. The actigraph resembles a wrist watch and measures the person's sleep.

The patient will usually be tested overnight in a medical center equipped with a polysomnograph. The polysomnograph is a machine that measures the electrical activity of the heart and brain, breathing, and eye movement while the patient is sleeping. Another sleep test is called the multiple sleep latency test or MSLT. The patient is given a chance to sleep every two hours during normal waking time. Observations are made of the time taken to reach various stages of sleep. The MSLT measures the degree of the patient's daytime sleepiness and also detects how soon REM sleep begins.

# Treatment

Treatment of narcolepsy involves a combination of medications, lifestyle changes, and psychotherapy. The most common medications given are stimulants that resemble amphetamine. It is important for persons taking stimulants to notify their employer, because the drugs may show up during pre-employment urine tests. Another type of drug that may be prescribed is a drug called Xyrem, which controls cataplexy and helps patients sleep better at night.

Lifestyle changes that are recommended for patients with narcolepsy include:

- Getting enough sleep at night. Patients are advised to go to bed and get up at the same time every day rather than changing their sleeping schedule frequently.
- Taking scheduled short naps at intervals during waking hours. This practice helps to reduce daytime sleepiness.
- Avoiding the use of alcohol and tobacco. These substances can make it harder to sleep at night.
- Getting regular exercise. Physical exercise four to five hours before bedtime helps many people with narcolepsy sleep better.
- Being careful to avoid driving when tired or sleepy.
- Joining a narcolepsy support group. Finding understanding and support from others with the same disorder is particularly helpful to patients who may have waited years to be diagnosed and suffered the loss of jobs, relationships, or educational opportunities in the meantime.

# Prognosis

Narcolepsy cannot be cured but it is not a fatal disease. People with narcolepsy can expect to live normal life spans (barring accidents). They can also expect to lead normally productive and meaningful lives with proper care and periodic consultations with a sleep specialist.

## Prevention

There is no way known to prevent narcolepsy.

# WORDS TO KNOW

- Automatic behavior: Activity that a person with narcolepsy can carry out while partially awake but is not conscious of at the time and cannot recall afterward.
- **Cataplexy:** Sudden loss of tone in the voluntary muscles.
- **Hypnagogic:** Referring to the period of partial alertness on the boundary between sleeping and waking.
- **Hypocretin:** A protein produced by certain brain cells that promotes wakefulness and helps to

regulate the sleep/wake cycle. It is also known as orexin.

- Non-rapid eye movement (NREM) sleep: The first phase of a sleep cycle, in which there is little or no eye movement.
- **Rapid eye movement (REM) sleep:** The phase of a sleep cycle in which dreaming occurs; it is characterized by rapid eye movements.
- **Sleep cycle:** A period of NREM sleep followed by a shorter phase of REM sleep. Most adults have four to six sleep cycles per night.

# The Future

New drugs that will help narcoleptic patients remain awake were at the clinical trial stage in late 2008. Some researchers in Germany were experimenting with a nasal spray containing hypocretin that might be beneficial to people with narcolepsy. Another possibility is the use of stem cell transplants to restore the cells in the brain that make hypocretin, but that approach will likely take several decades to be successful.

SEE ALSO Restless legs syndrome; Sleep apnea

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# Necrotizing Fasciitis

# Definition

Necrotizing fasciitis (NF) is a rare but potentially deadly infection of the deeper layers of the skin and the fascia, which is a sheet of connective tissue that overlies the muscles, bones, blood vessels, and internal organs. The fascia protects these parts of the body, provides them with structure, and acts as a shock absorber. Destruction of the fascia is dangerous because this sheet of tissue helps the body repair itself and acts as a protection against infection.

# Description

Necrotizing fasciitis is a life-threatening infection of the skin and fascia that develops when certain types of bacteria get into the body through breaks in the skin. These may be accidental cuts or scrapes or the result of surgery. "Flesh-eating bacteria" is a bit of a misnomer because the germs do not literally eat the soft tissue. Instead they secrete a substance that rapidly kills the tissue.

NF is particularly dangerous because it is often misdiagnosed until it is too late for the patient to survive. The early symptoms of necrotizing fasciitis often mimic the symptoms of influenza, a pulled muscle, or arthritis. In the case of necrotizing fasciitis following surgery, the patient's pain is often misdiagnosed as normal postoperative discomfort.

#### Also Known As

Flesh-eating disease, fasciitis necroticans, soft tissue gangrene

#### Cause

Bacteria, most commonly Group A streptococci

#### Symptoms

Rapidly spreading skin swelling, severe pain, smelly discharge, tissue death

Duration

Hours to days

#### Necrotizing Fasciitis

Scarring on a man's arm from an attack of necrotizing fasciitis. AP IMAGES.



# **Demographics**

Necrotizing fasciitis was first reported in a medical publication in 1848. The Centers for Disease Control and Prevention (CDC) estimates that there are between 500 and 800 cases each year in the United States. Most affect adults; the average age of patients is between thirty-eight and forty-four years. Males are three times more likely to be affected. About half of all patients diagnosed with NF were previously strong and healthy.

Some people, however, are at greater risk of developing NF:

- People with diabetes, heart disease, and other disorders that affect blood circulation
- Drug addicts and alcoholics
- People with weakened immune systems, including those who have received organ transplants
- People with HIV infection

# **Causes and Symptoms**

The cause of NF is infection by bacteria that enter the body through cuts or other breaks in the tissue, or infection of an incision after surgery. In some cases the injury may not even be apparent to the patient; one case in 2005 involved a firefighter who died from NF after spending several hours in bacteria-contaminated floodwaters rescuing trapped people. Most cases of NF involving injuries to the extremities are caused by Group A streptococci; these are sometimes called GAS infections. NF following surgery can be caused by a variety of other bacteria, sometimes several species at the same time.

Once in the body, the bacteria spread along the fascia, secreting toxins that destroy the fascia, the deeper layers of the skin, and muscle tissue. The toxins also prevent the flow of blood to the damaged tissue. As the infection spreads, the tissue reddens and swells. The speed of the spread depends on the depth of the infection within the fascia. Some doctors divide the symptoms of NF into three phases:

- Early (first twenty-four hours): Pain in the area of the cut or injury that quickly becomes more severe than would be expected; flu-like symptoms (diarrhea, nausea, vomiting, fever, confusion, dizziness, and weakness); rapid dehydration.
- Advanced (next three to four days): The affected body part swells and develops a purplish rash; blisters form and fill with a blackish fluid; the injured tissue starts to look decayed or dead (may be bluish or white).

# A Survivor's Story

On Monday, June 12, 2006, I had what I thought to be a nasty bout of the flu.... By Wednesday evening I was vomiting every hour and could not catch my breath in between. I woke my husband and told him I think we better go to emergency.... We drove the 19 miles (30 kilometers) to our local hospital [in Canada] and I was admitted immediately. The original diagnosis was diabetic acid ketosis and after a week things had stabilized except for a persisting high temperature and a small lesion on the inside of my left groin. On June 21 I was told I would be transferred by ambulance to a major city's hospital (three hours away by car) for further testing....

The next day I fully remember is August 27, 2006. By 7:00 p.m. that night the hospital called my husband and told him he needed to come right now because I probably would not live.... I was only given a 30 percent chance of surviving and less than 5 percent chance I would keep my left leg. The necrotizing fasciitis had spread down my leg to the knee, across my back and up to my shoulder blades....

After sixteen surgeries I am happy to say that I have my leg, though severely damaged and extreme scarring from several skin grafting sites and skin donor sites. In total I spent six months in hospital, two and a half months in the intensive care unit and then transferred to a burn unit.... At the end of October 2006 I was finally transferred to a rehabilitation hospital where I was able to get the help to learn how to walk again. On December 6, 2006, I was able to come home. Once home I was only able to use a walker and wheelchair. By March I was walking with only the use of a cane and in April I had my last reconstructive surgery. My life has changed so much.... I can now almost do everything the way I used to. Walking will always be an issue but I am now able to drive and am back to work part-time.

• Critical (next two days): The person's blood pressure drops rapidly, the patient loses consciousness, and the entire body goes into toxic shock.

#### Diagnosis

Doctors diagnose necrotizing fasciitis on the basis of the appearance of the injured tissue, the patient's other symptoms, and a blood test to identify the organism(s) causing the infection. In some cases the doctor can diagnose NF simply by passing a gloved hand between the tissue layers in the injured area; this procedure could not be done with healthy fascia. In some cases a surgeon may remove a small amount of the dying tissue for examination under a microscope. Treatment typically begins even before the bacterium is identified because speed of treatment is essential.

## Treatment

Patients with NF are given antibiotics intravenously, often a combination of two or three antibiotics to kill all the possible bacteria that may be involved in the infection. The patient is also examined by a surgeon, who will remove all the dead and infected tissue. This process is called debridement, and will be repeated until the surgeon is sure that all the infected tissue has been cut away. It is critical to remove this tissue to prevent the infection from spreading farther. In some cases the surgeon will have to amputate a limb to prevent death, and skin grafts may be necessary afterward.

Most patients will need to be monitored in an intensive care unit for several days or even several weeks. Some patients with NF are given additional treatment in a hyperbaric oxygen chamber. This is a special room in which the patient is treated with large amounts of oxygen.

## Prognosis

The prognosis for necrotizing fasciitis depends on the promptness of treatment, the virulence (strength) of the organism causing the infection, and the patient's overall health. Without speedy treatment, between 70 and 75 percent of patients will die. The death rate is about 25 percent even in patients who were diagnosed quickly and treated at once.

Patients who survive may take months to years to recover completely. It is not unusual for people to require several operations, treatment in a burn center or other specialized hospital, a long period of rehabilitation, plastic surgery to restore normal appearance, and psychotherapy to deal with the depression that often follows NF.

#### Prevention

There is no way to completely prevent NF. The best way to lower one's risk of NF is to keep the skin intact as much as possible and clean the skin thoroughly after a cut, scrape, or similar injury. Applying an antibacterial ointment to open cuts or sores also helps. Last, people who have strep throat should cover their mouth when they cough and dispose promptly of soiled tissues or handkerchiefs, as the bacterium that causes strep throat is a common cause of NF as well.

#### **The Future**

NF may become more common in the future, partly because there are more people with organ transplants, HIV infection, diabetes, and other conditions that put them at increased risk of NF.

SEE ALSO Staph infection; Strep throat; Toxic shock syndrome

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#### WORDS TO KNOW

**Debridement:** The medical term for the surgical removal of dead or damaged soft tissue.

**Fascia:** A sheet of connective tissue that covers and binds together the muscles and other internal body structures.

**Gangrene:** Decay and death of soft tissue due to loss of blood supply.

**Necrotizing:** Causing the death of soft tissue.

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# 0



#### Genetic

Infection

Injury

Multiple

Other

Unknown

Also Known As Overweight, being fat

#### Cause

Genetic, behavioral, and environmental factors

#### Symptoms

Body mass index (BMI) above 30; waist measurement greater than recommended

#### Duration

Years



#### Definition

Obesity is defined as a condition in which a person has accumulated so much adipose (fatty) tissue that his or her health is impaired. It is also defined by a person's body mass index (BMI) or waist measurement. The BMI is an indirect measurement of body fat. To calculate an adult's BMI in English measurements, multiply a person's weight in pounds by 703.1, and divide that number by the person's height in inches squared. A person with a BMI above 25 is considered overweight; a BMI over 30 is considered obese; and a BMI above 40 is considered morbid obesity. In terms of waist measurement, women with a waistline greater than 35 inches (88.9 centimeters) and men with a waistline greater than 40 inches (101.6 centimeters) are considered obese.

Some doctors think that obesity itself should be defined as a disease, while others maintain that it should be considered more broadly as an unhealthy condition that increases a person's risk of diabetes and other diseases.

#### Description

Obesity is a condition that develops when a person takes in more food calories than his or her body burns up in the course of one's daily activities. The body turns those excess calories into fat. Overweight refers to an

#### Obesity

Two normal mice and one with a gene for obesity. The obese mouse is a result of genetic research. AP IMAGES.



excess of body weight compared to set standards. The extra weight may come from muscle, bone, fat, or body water. Obesity refers specifically to having an abnormally high proportion of body fat. Thus it is possible for an athlete to be overweight without being obese if his or her extra weight consists of muscle rather than fat tissue.

Obesity is a concern to doctors because it is associated with a number of diseases and disorders that can shorten a person's life expectancy as well as lower one's quality of life. Some of these diseases and other health problems include:

- Type 2 (adult-onset) diabetes
- High blood cholesterol levels
- Heart disease
- Stroke
- High blood pressure
- Osteoarthritis
- Sleep apnea
- Gallbladder disease
- Complications of pregnancy
- Depression
- · Increased risk of breast, colorectal, or kidney cancer
- Increased risk of complications following surgery

#### Demographics

Obesity is a growing health problem in all countries in the developed world. The World Health Organization (WHO) estimated in 2005 that 10 percent of the world's population, or 400 million people, were obese, with another 760 million overweight. In the United States, about twothirds of adults are overweight compared to the ideal weight for their sex, age, and height; one-third of American adults are obese. The proportion of obese adults in the general American population has more than doubled since 1960. Obesity costs the United States about \$100 billion each year, \$52 billion for direct health care costs and \$33 billion for weight-loss products or programs.

Obesity is equally likely to affect men and women. There are, however, racial and ethnic differences within the American population. Groups that have a higher than average risk of obesity include the Pima Indians of Arizona and several other Native American tribes; Hispanics; African Americans; and Pacific Islanders.

Obesity is often associated with eating disorders in adults; about 30 percent of obese adults have a history of bulimia or binge eating.

#### **Causes and Symptoms**

Obesity is thought to be the end result of a combination of factors rather than having a single cause. Although obesity can be briefly described as an imbalance between food energy taken in and energy used up in exercise, eating and exercise are intertwined with other factors in complicated ways.

- Genetic factors. Researchers think that these account for 40–70 percent of the variations in human body size. Genetic factors affect how efficiently a person's body burns food and where on the body the excess fat is stored. People whose fat is stored around the abdomen have a higher risk of health problems than those whose extra weight is carried on the hips.
- Family environment. A person whose parents eat a lot of highcalorie foods is likely to adopt their eating habits.
- Age. People's bodies lose muscle tissue and gain fat as they age, and their calorie requirements drop; thus people who may not have been obese as young adults may become obese as they grow older.
- Sex. Men have more muscle tissue and less fat than women, and their calorie requirements for maintaining their body weight are higher than women's. Thus women are more likely than men of

the same height and weight to gain weight if they consume the same number of calories as the men.

- Medical conditions. People with Cushing's syndrome, disorders of the thyroid gland, or depression are at increased risk of obesity.
- Medication side effects. People who must take corticosteroid drugs, antiseizure medications, or antidepressants may gain weight on these drugs.
- Emotional factors. Some people use food to soothe their feelings when they are sad, angry, bored, or upset.
- Quitting smoking. People who quit smoking often gain weight because food tastes better after they quit. In addition, the nicotine in tobacco raises the rate at which the body burns calories, so the former smoker's body needs fewer calories to maintain its weight.
- Pregnancy. Most women retain 4 to 6 pounds (1.8 to 2.7 kilograms) after each pregnancy; over time, these weight gains can lead to obesity.

#### Diagnosis

The diagnosis of obesity in adults may be based on the CDC body mass index tables, or it may be based on other measurements. One common test involves measuring the thickness of the skin fold over the triceps muscle on the upper arm, although this measurement may not be accurate unless performed by a trained technician. Another test involves measuring the person's waist circumference at its widest point, usually at or just below the belly button.

#### Treatment

Treatment of obesity is usually more complicated than simply placing people on a calorie-restricted diet or telling them to get more exercise. The doctor must take into account other health conditions that may affect the patients, their medications, occupation, and other lifestyle factors. For example, a person who lives alone can control food purchases or daily workouts more easily than someone who has to take the eating habits and time schedules of other family members into consideration. Similarly, someone with osteoarthritis may need to find forms of exercise that will not damage sore joints. In some cases the doctor and patient can work out a diet and exercise plan with the help of a registered dietitian or physical therapist, so that the patient can eat a nutritious diet that includes one's favorite foods and find a form of exercise that he or she likes as well. One of the keys to sticking with a weight-loss program is satisfaction; people who feel deprived of the foods they like or bored by their physical exercise program are likely to quit.

It helps to remember that losing even a small amount of weight can be beneficial. Some doctors call this approach the "10 percent solution"—that is, losing just 10 percent of one's excess weight can lower the risk of heart disease and diabetes. If the person is able to keep that 10 percent weight loss for six months or longer, he or she can consider losing more weight.

People who have been unable to lose weight by a combination of exercise and food intake, or who are morbidly obese, may be treated by medications or weight-loss surgery. The two major medications that have been approved by the Food and Drug Administration (FDA) for weight loss are Meridia, which works by making the person feel full more quickly during a meal, and Xenical, which works by lowering the amount of fat that the body absorbs from the intestines. Both drugs have side effects, and weight loss tends to be modest, about 13 pounds (5.9 kilograms) in a year.

Weight-loss surgery, known as bariatric surgery, is usually limited to people with a BMI over 40 or those with a BMI over 35 combined with type 2 diabetes, sleep apnea, or heart disease. There are two basic types of surgery for weight loss, restrictive and malabsorptive. Restrictive procedures work by closing off part of the stomach so that the person cannot eat as much without feeling full. In malabsorptive procedures, the surgeon creates a bypass around the part of the small intestine where most of the calories in the food are absorbed. This type of surgery lowers the amount of calories that the body absorbs as well as lowering the patient's food intake. Bariatric surgery is not for everyone; it can produce serious complications that include bloating, nausea, and diarrhea. In addition, the patient will need to see the doctor periodically for the rest of his or her life.

#### Prognosis

According to the Centers for Disease Control and Prevention (CDC), obese adults have a 10–50 percent increase in their risk of dying from any cause, compared to people whose weight is normal or only slightly overweight. Obesity shortens a person's life expectancy by six to seven years on average, and accounts for 112,000 excess deaths each year in the United States compared to people of normal weight. Men with a BMI over 40 have their life expectancy shortened by twenty years, and women by five years.

#### WORDS TO KNOW

Adipose tissue: Fatty tissue.

- **Binge:** An episode of eating in which a person consumes a larger amount of food within a limited period of time than most people would eat in similar circumstances.
- **Body mass index:** BMI. An indirect measurement of the amount of body fat. The BMI of adults is calculated in English measurements by

multiplying a person's weight in pounds by 703.1, and dividing that number by the person's height in inches squared.

- **Cushing syndrome:** A disorder caused by the excess secretion of cortisol by the pituitary gland.
- **Ideal weight:** Weight corresponding to the lowest death rate for individuals of a specific height, gender, and age.

#### Prevention

Preventing weight gain or maintaining weight loss can be difficult in a society where inexpensive high-calorie snacks make it easy to overeat and television or video games make it easy to avoid physical activity. The following tips, however, can help a person stay committed to a weight-loss or weight-maintenance program:

- Make time each day—thirty to sixty minutes—for fast walking, swimming, or other forms of moderate exercise.
- Build a daily menu around fruits, vegetables, and whole-grain foods rather than sweets or snacks that are high in fat.
- Learn to recognize the situations or triggers that lead to overeating. One way to spot these patterns is to keep a food diary.
- If necessary, get help for depression or other psychological problems related to overeating.
- Keep track of weight and waist size on a regular basis.

#### The Future

It is likely that obesity will increase in the future, not just in the United States but worldwide. The World Health Organization (WHO) reported in 2006 that the only part of the world where obesity is still unusual is sub-Saharan Africa. Obesity is rising in all age groups, including the elderly, as well as in almost all countries.

Since the discovery in 1994 of leptin, a hormone that regulates the brain's sense that the body has had enough to eat, researchers are

studying it in hopes of learning more about the biological factors that influence appetite and food intake in humans. Other researchers are looking at the emotional and social factors that encourage people to overeat, and to test the effectiveness of psychotherapy as well as medications in weight-loss programs.

**SEE ALSO** Bulimia; Childhood obesity; Diabetes; Hypercholesterolemia; Hypertension; Sleep apnea

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## Obesity, Childhood

See Childhood obesity.

# m

### **Obsessive-Compulsive Disorder**

#### Definition

Obsessive-compulsive disorder (OCD) is an anxiety disorder. People with OCD experience recurrent bothersome thoughts or mental images (obsessions) that worry the person. He or she then tries to control or ward off the anxiety by carrying out repetitious behaviors or mental acts (compulsions).

#### Description

OCD is sometimes described as a malfunction of the brain's information processing system. Everyone has upsetting thoughts or impulses from time to time—such as the urge to shout dirty words in public or thinking about hitting someone. However, most people are able to let go of these things and not worry about them. A person with OCD gets stuck on the thoughts or impulses and cannot put them aside. These thoughts or impulses are called obsessions. The person who has them may think that he or she is "going crazy" or will not be able to keep from acting on the thoughts. To cope with the anxiety, the person with OCD engages in repetitive behaviors or mental acts to undo, counteract, or control the obsessions. These behaviors are called compulsions.

Common obsessions include fears of contamination by germs or dirt; fear that one has harmed someone; thoughts of violence or of killing a pet or family member; worrying about thoughts that violate one's religious beliefs; and fear of performing sexual acts that the person dislikes. Common compulsions include repeated hand washing or bathing; checking doors or car windows over and over to be sure they are locked; counting objects; insisting that personal possessions like clothes in a closet or items on a desk be arranged "just so"; touching objects in a specific sequence; and hoarding items that are not needed.

It is important to recognize that people with OCD are distressed by these behaviors and usually realize that they do not make sense. Compulsions may consume several hours of the patient's day, interfering with work, family life, and other activities. They can also be harmful to health. There are instances of people damaging their skin by frequent hand washing or taking long showers. Unlike people with substance abuse or

#### Also Known As OCD

#### Cause

Biological vulnerability, learned behavior, changes in brain chemistry

#### Symptoms

Recurrent thoughts or impulses; repeated acts to control anxiety caused by the thoughts

#### Duration

Years; may begin in childhood



Brain images of a person with obsessive-compulsive disorder. Red and yellow areas show increased activity with an increase in symptoms. WDCN / UNIV. COLLEGE LONDON / PHOTO RESEARCHERS, INC.

eating disorders, people with OCD do not find their rituals pleasurable or satisfying; the acts are done only to manage their fears.

#### **Demographics**

OCD is a common anxiety disorder. According to some estimates, one adult in every fifty currently has the disorder, and two out of fifty have had it at some point in their lives. As of 2008 it is thought that about 2.2 million adults in the United States have OCD. It is likely, however, that the disorder is underdiagnosed because many people who suffer from it are embarrassed by their symptoms and often skilled at hiding them from others.

OCD can begin at any age, including childhood. In fact, between a third and a half of adults diagnosed with the disorder say that their symptoms began in childhood. The most common age for the emergence of symptoms is between ten and twenty-four years.

OCD is equally common in all races and ethnic groups in the United States, although patients' specific obsessions are sometimes influenced by their cultural background. For example, fears of violating religious practices or beliefs are reported to be more common among Roman Catholics and Orthodox Jews than among Protestants.

Males and females are equally likely to develop OCD. However, males are more likely to begin showing symptoms in childhood and

#### Howard Hughes and OCD

During his lifetime, Howard Hughes (1905–1976) was an aviator, film producer, and one of the wealthiest people in the world. In 1953 he founded the Howard Hughes Medical Institute, which is one of the largest privately funded medical research institutions in the United States today. Although the institute has helped many people with its research on genetics and the immune system, it could not help Hughes with his OCD.

Hughes's behavior was fairly normal until the 1930s. At that time, his friends and business partners began to notice some strange behaviors, such as a compulsion to sort peas by size with a fork for hours. Another of Hughes's obsessions concerned contamination. He used paper tissues to pick up objects to avoid being exposed to germs. He also burned his clothing if he found out that someone he had met while wearing it had an illness. In the late 1960s, his obsession with contamination focused on radioactivity. At the time, Hughes was living in Nevada, where underground nuclear weapons tests were being conducted. He was so worried about being poisoned by radiation that he offered two successive presidents-Lyndon Johnson and Richard Nixonmillion-dollar bribes to stop the tests.



Howard Hughes. COURTESY OF THE LIBRARY OF CONGRESS.

Hughes eventually left the United States for the Bahamas. When he died in 1976, he weighed only 90 pounds and could only be identified by his fingerprints. The cause of death was kidney failure brought on by malnutrition. Howard Hughes's OCD led him to fear that his food was poisoned.

adolescence, and females are more likely to develop symptoms in their early twenties.

#### **Causes and Symptoms**

At one time it was thought that OCD was caused by childrearing practices that made the person anxious. Such compulsions as repeated washing or checking door locks were explained as rituals intended to please parents who were overly concerned with cleanliness or safety. This type of psychological explanation is no longer considered useful. There are several recent theories about the possible causes of OCD:

- Genetic. It is known that having other family members with OCD increases a person's risk of developing the disorder. Although two genes have been linked to OCD, one discovered in 1994 and the other in 2007, researchers have not yet been able to prove that either or both cause the disorder.
- Abnormally low levels of serotonin in the brain. Serotonin is a chemical produced by the brain that regulates mood, appetite, sleep, and memory. One piece of evidence that supports the serotonin theory is that patients with OCD, who are given a type of antide-pressant that makes more serotonin available to brain cells, obtain some relief from their symptoms. These antidepressants are called selective serotonin reuptake inhibitors, or SSRIs.
- Differences in brain structure. Researchers who have used magnetic resonance imaging (MRI) to map the regions of the brain have found that people with OCD have abnormally high levels of activity in some areas.
- Infections. OCD in children has sometimes been attributed to a complication of strep throat. This theory holds that the child's body produces antibodies against the strep throat bacteria. These antibodies then attack the brain and cause a sudden onset of OCD. However, this theory is controversial and is considered unproven.

The symptoms of OCD are obsessions and compulsions that interfere with the person's schoolwork, job, or social functioning, and that have no real-life basis in the person's present situation. (For example, a person with an immune system disorder would have a real-life reason for worrying about germs and infection, and a person living in a high-crime neighborhood would be understandably very concerned about checking locks). Common obsessions include:

- Fear of dirt, germs, radioactivity, or other types of contamination
- Doubting whether one has completed a task
- Needing to have things in perfect order or in some kind of symmetrical arrangement
- Sexual thoughts
- Aggressive or violent thoughts.

Common compulsions include:

- Repeatedly washing one's body, clothing, or other personal items
- Repeatedly checking one's work
- Constantly asking others for reassurance
- Repeatedly counting or rearranging items
- Counting numbers in certain patterns (all odd or even numbers, for example)
- Hoarding (such as buying several years worth of cleaning supplies) or being unable to throw out old magazines or worn-out items

#### Diagnosis

The diagnosis of OCD is often delayed because patients are ashamed of their symptoms and skilled at hiding them. It has been estimated that it takes an average of seventeen years from the time that a patient's symptoms begin for them to be diagnosed correctly and receive treatment for OCD. Another reason for the delay is that a person with OCD often has other disorders, including substance abuse disorders, bipolar disorder, panic disorder, or depression, and the OCD symptoms may be attributed to the other disorders. In children, OCD is sometimes misdiagnosed as autism or Tourette syndrome.

There are no laboratory tests for OCD. The person's primary care doctor will refer the patient to a psychiatrist or psychologist for a specialized interview. The diagnosis is based on a combination of the patient's history of symptoms and his or her answers to a diagnostic questionnaire. The questionnaire most often used is the Yale-Brown Obsessive Compulsive Scale (Y-BOCS). The Y-BOCS has ten items, five for obsessions and five for compulsions. The questions evaluate the time consumed by symptoms, the extent to which they interfere with functioning, how much they distress the patient, and what the patient has done to try to control them.

#### Treatment

Treatment for OCD is usually based on a combination of medications and a type of psychotherapy called exposure and ritual prevention (or exposure and response prevention) or ERP. ERP is a form of cognitive behavior therapy in which patients are forced to confront their fears without resorting to their usual safety rituals. For example, someone who is afraid of contamination might be asked to touch an object that has been touched by another object that has been touched by a piece of cloth from a "contaminated" location or source, and then refrain from washing his or her hands. The next time the patient might be asked to touch an object that has been directly touched by the cloth, and again not to wash. In most cases the patient's anxiety level drops fairly quickly and he or she can then give up the safety ritual.

The medications usually prescribed to treat OCD are the selective serotonin reuptake inhibitors or SSRIs. These include drugs like Paxil, Prozac, and Zoloft. As noted earlier, these drugs work by increasing the amount of serotonin available to some of the nerve endings in the brain.

A very small number of patients with severe OCD that does not respond to medications or ERP are treated surgically. The surgeon makes a small cut in a part of the brain called the cingulate bundle. This technique produces significant benefits for about 30 percent of patients who receive the operation. It is considered a treatment of last resort for severe OCD.

#### Prognosis

The prognosis of OCD varies from person to person. The disorder rarely goes away on its own. About 70 percent of patients benefit from treatment. However, the patient's symptoms may increase and decrease in a cyclical pattern over time. About 15 percent of patients get steadily worse over time even with treatment and may eventually become unable to function.

#### Prevention

There is no known way to prevent OCD. However, early diagnosis and prompt treatment can help to prevent the patient's symptoms from getting worse.

#### The Future

It is not known at present whether OCD is becoming more commonplace in the general population or whether a recent increase in the number of reported cases is due to wider recognition of the disorder and improved diagnosis.

Researchers are presently studying the effectiveness of newer drugs in treating OCD as well as deep brain stimulation or DBS. In DBS, thin wires are implanted in the parts of the brain that have been linked to OCD symptoms. A battery-powered stimulator sends electrical pulses to the

#### WORDS TO KNOW

**Compulsion:** A repeated behavior or mental act carried out to control or neutralize obsessions.

**Obsession:** A recurrent, distressing, intrusive thought, image, or impulse.

Selective serotonin reuptake inhibitors (SSRIs): A group of antidepressants that work by increasing

the amount of serotonin available to nerve cells in the brain.

**Serotonin:** A brain chemical that influences mood, anger, anxiety, body temperature, and appetite. It may be involved in OCD.

brain at regular intervals in order to interfere with the activity of the nerve cells in the target areas. DBS has already been used to treat Parkinson disease and is considered an experimental treatment for Tourette syndrome.

**SEE ALSO** Bipolar disorder; Depression; Panic disorder; Strep throat; Tourette syndrome

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## **m** Osteoarthritis

#### Definition

Osteoarthritis, or OA, is a disease of the joints in which the cartilage that cushions the bones in the joints starts to break down and wear away. As the cartilage disappears, the bones begin to rub against each other, causing pain, stiffness, loss of flexibility and mobility in the affected joints, and the formation of bony spurs around the joints.

OA can be classified as either primary or secondary. Primary osteoarthritis occurs without an injury to the joint or other obvious cause. Secondary OA develops as a result of another disease or injury to the joint.

#### Description

Osteoarthritis is a disorder of the joints in which the gradual wearing away of the cartilage within joints leads to pain, stiffness, and loss of the ability to move the joint freely. OA can affect almost any joint in the body, although it is most commonly found in the large weight-bearing joints—the hips, knees, spine, and feet—and in the hands and fingers. Although osteoarthritis is not caused by the aging process itself, it is largely a disease of older adults. In most cases the disorder comes on slowly and gradually over a period of years rather than appearing suddenly.

Osteoarthritis differs from rheumatoid arthritis (RA) in that it does not affect organs or body systems other than the joints. Although OA may be painful, it does not affect the blood vessels, digestive system, lungs, or other internal organs. Fever or redness in the joints is more likely to indicate RA than osteoarthritis.

It is possible for people to have osteoarthritis without noticeable pain or swelling. For example, some doctors estimate that 80 percent of adults

#### Also Known As

OA, degenerative joint disease, osteoarthrosis

#### Cause

Damage to joints resulting from low-grade inflammation and loss of cartilage

#### Symptoms

Pain and swelling in weight-bearing joints; morning stiffness; difficulty moving the joint

#### Duration Years

#### Osteoarthritis

Hands of an elderly person deformed by osteoarthritis. © MEDICAL-ON-LINE / ALAMY.



over sixty-five have evidence of OA when they are x-rayed, but only 60 percent of this group have joint pain and other troublesome symptoms of the disease.

The severity of osteoarthritis varies from patient to patient. Some people are only mildly affected and can manage their symptoms with over-the-counter pain relievers when needed. Others, however, find their lives disrupted by the disease. They may have to change occupations, become completely unable to work, or lose their ability to carry out ordinary daily activities. Elderly adults with severe OA may no longer be able to live independently.

#### Demographics

Osteoarthritis is the most common joint disease among older adults in the United States. It affects nearly 21 million people in the United States and accounts for 25 percent of visits to primary care physicians. Half of all prescriptions for nonsteroidal anti-inflammatory drugs (NSAIDs) are written for patients with OA.

The rate of OA in the population increases with age; it affects about 30 percent of people over forty-five but almost 80 percent of people over eighty. Osteoarthritis is equally common in men and women between the ages of forty-five and fifty-five. However, more women than men are affected in the over-fifty-five age group. Younger adults, however, can

develop OA as a result of repeated injuries to their joints, abnormally shaped joints, or genetic disorders that affect the joints.

Some people are at increased risk of osteoarthritis:

- People who are severely overweight. Obesity increases the strain on such weightbearing joints as the knees, lower spine, and hips.
- People from families with a history of OA. Some researchers think that genetic factors are involved in as many as 60 percent of OA cases, although no specific genes have been linked with OA currently. Women are more likely than men to be affected by a family history of osteoarthritis in the hands and fingers.
- People with misshapen joints or defective cartilage. Structural abnormalities in a person's joints increase the risk of OA.
- Race and ethnicity. African American women are at increased risk of developing osteoarthritis of the knee, although the reasons for this difference are not completely understood.

#### Coping with Osteoarthritis

One of the most effective forms of self-care in coping with OA is keeping a positive attitude. In addition to watching their weight and getting a good night's sleep, people with OA can help themselves by:

- Focusing on what they can do rather than what they cannot.
- Breaking down daily chores and activities into smaller tasks.
- Looking at ways to minimize emotional stress as well as stress on their joints.
- Balancing rest with activity; keeping up moderate levels of exercise without overdoing.
- Practicing meditation, relaxation techniques, or guided imagery to manage pain.
- Putting together a support community of friends, family members, and health care professionals.
- Learning to listen to their body's signals and recognize their limits.

- Pregnant women.
- People who have injured a joint in an accident or participation in sports that puts pressure on the joints (running, martial arts, contact sports, tennis, baseball, etc.)
- People who have been diagnosed with other diseases that affect the joints, such as gout, Lyme disease, rheumatoid arthritis, or Paget disease.

#### **Causes and Symptoms**

Primary osteoarthritis is caused by a gradual loss of water from the cartilage in the joints. Healthy cartilage contains between 65 and 80 percent water, which allows it to protect the bones against the effects of gravity on the body and to keep the joints moving smoothly. The bones in the joints and the cartilage that surrounds them are encased in a structure called a joint capsule. The joint capsule in turn is lined with a membrane that secretes a fluid that lubricates the joints. As people age and their cartilage shrinks from loss of water, the ends of the bones begin to produce outgrowths known as osteophytes or spurs. As the cartilage wears away, it releases breakdown products into the fluid produced within the joint capsule, resulting in a mild inflammation and swelling of the joint. The swelling in turn stretches the joint capsule, leading to the pain and stiffness associated with OA.

In secondary OA, the gradual loss of cartilage is triggered by a traumatic injury or infectious disease. The disease progresses in the same way as in primary osteoarthritis.

The symptoms of OA typically include:

- Pain in the joint during use, especially when used after a period of inactivity
- Soreness in the joint when light pressure is applied
- Stiffness in the joint, usually worse in the morning
- Cracking, popping, or grating sensation or sound when the joint is moved or used
- Swelling of the tissues around the joint
- Hard bumps or lumps around the joint caused by bone spurs
- Discomfort in the joint that gets worse during rainy or humid weather

#### Diagnosis

Pain is the symptom of OA that is most likely to bring patients to their doctor. The doctor will begin by taking a history of the symptoms, including a family history of osteoarthritis. The doctor may ask whether the symptoms are affected by the type of activity, level of activity, or weather conditions.

The next step in diagnosing osteoarthritis is a physical examination of the affected joints. The doctor may press on the joints, feel the area around the joints for bony lumps, or try to move the joints to see whether their range of motion is limited.

Laboratory tests and imaging studies may include a blood test to rule out rheumatoid arthritis; an x-ray of the affected joint; withdrawal of a small amount of fluid from the affected joint to rule out gout or an infection; or the insertion of a miniature camera to look inside the joint. This type of examination is called arthroscopy.

#### Treatment

There is no cure for OA. Therapy is aimed at relieving pain and swelling, and keeping the joint functional. Treatment of osteoarthritis begins with a combination of lifestyle modifications, mild pain relievers, and exercises or physical therapy. Overweight patients are encouraged to lose weight to reduce stress on the weight-bearing joints of the body. In some cases, the patient may be advised to correct poor posture, which can also place stress on the joints. Muscle-strengthening exercises can help to keep the joints flexible. Physical therapy may include exercising in a pool, lowimpact stretching exercises, or other exercises tailored to the individual by a licensed physical therapist.

Applying heat or cold to the affected joints is helpful for many patients. Using hot packs for twenty minutes several times a day, sitting in a warm (not hot) bath, or using a heating pad relieves stiffness, while applying ice packs relieves muscle spasms. Some people are also helped by therapeutic massage, provided that the therapist understands the symptoms of osteoarthritis and does not put too much pressure on the affected joints.

There are several types of medications that can be used to relieve the pain of OA:

- Tylenol. Tylenol (acetaminophen) is a pain reliever that has few side effects and works well for patients with only mild symptoms of OA.
- NSAIDs. These medications reduce inflammation as well as relieve discomfort. Many are available over the counter, including Motrin, Advil, and Aleve. NSAIDs may, however, upset the stomach or cause stomach ulcers and kidney problems when used in high doses over a long period of time. Patients should check with their doctor about long-term use of NSAIDs.
- Cortisone injections. These are given for relief of severe joint pain. They should not be used more than four times a year, however, as they can cause long-term damage to the joints.
- Tramadol. Tramadol is a stronger pain reliever than NSAIDs but carries the risk of addiction. It requires a doctor's prescription.

- Viscosupplements. These are drugs resembling a compound that is normally found in joint fluid. They can be used only for osteoarthritis of the knee. The doctor injects the supplement into the knee joint over a period of three to five weeks. Viscosupplements are effective for about six months.
- Glucosamine and chondroitin sulfate. These are dietary supplements that some doctors recommend for joint pain. There is some evidence that these supplements can help control the pain of OA, although they do not seem to grow new cartilage.

Severe osteoarthritis may require surgical treatment. Procedures that have been done to treat OA include:

- Joint replacement. This type of surgery is usually done for hip and knee joints. The damaged joint is removed and replaced with an artificial joint made of plastic, metal, or ceramic parts.
- Arthroscopy. Surgeons can use an arthroscope to remove torn and damaged cartilage or wash out a joint as well as look inside the joint.
- Realignment of a damaged or misshapen bone to relieve stress on an arthritic joint.
- Bone fusion. Surgeons can fuse bones together in a damaged joint to relieve the pain of arthritis. This procedure reduces the joint's flexibility, however.

#### Prognosis

The prognosis of osteoarthritis varies. It depends on the patient's age, weight, general health, and the specific joints that are affected.

#### Prevention

Keeping one's weight within recommended limits for one's age, gender, and height is a good preventive measure, as is keeping physically fit.

#### The Future

Osteoarthritis has been regarded for decades as a disorder produced by simple wear and tear on the body's joints. More recent research indicates, however, that arthritic joints produce abnormal chemicals that lead to the breakdown of cartilage. Geneticists are now looking for specific genes that may be involved in the production of these chemicals. If such genes

#### WORDS TO KNOW

- **Arthroscopy:** The use of a small device called an arthroscope to look inside and diagnose or treat an arthritic joint.
- **Cartilage:** A type of dense connective tissue that serves to cushion bones within joints.
- **Gout:** A disorder of the large toe or other joints caused by deposits of uric acid crystals in the affected joint.
- **Osteophyte:** A bony outgrowth or spur that develops in a joint affected by osteoarthritis. Osteophytes usually cause pain and limit the motion of the joint.
- **Paget disease:** A chronic disorder caused by a slow virus infection that results in deformed or enlarged bones.

are identified, scientists may be able to find ways to block their production of cartilage-destroying substances. Another area of research involves the study of doxycycline, an antibiotic used to treat sinus infections and acne, which may also be effective in slowing the destruction of cartilage in arthritic joints.

SEE ALSO Lyme disease; Obesity; Rheumatoid arthritis

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### Osteoporosis

#### Definition

Osteoporosis is a bone disease. Patients experience loss of bone mineral density followed by loss of bone strength.

#### Description

Osteoporosis is a disease that has no noticeable symptoms until the weakening of the bones leads to problems with posture, lower back pain, and brittle or easily broken bones. Although osteoporosis can appear at any age, it is most commonly a disease of adults. It develops when the wearing-out and removal of old bone—a process known as resorption outpaces the production of new bone tissue.

In most people, the bones become stronger and heavier during childhood and adolescence until they reach their maximum density and strength (peak bone mass) around age thirty. After that point, the bones gradually lose their density. People's peak bone mass and the rate at which they lose it in later life affects their risk of developing osteoporosis. The higher the peak bone mass at age thirty, the lower the risk of osteoporosis later in life.

Doctors divide osteoporosis into three categories or types. Types 1 and 2 are considered primary because they are not caused by other diseases or conditions. Type 3 osteoporosis is sometimes called secondary

#### Also Known As

Thin bones, brittle bone syndrome

#### Cause

Loss of calcium and other minerals from bone tissue

#### Symptoms

Back pain; loss of height; stooped posture; easily broken wrists, hips, or other bones

#### Duration

Years



Illustration of normal bone tissue versus that affected by osteoporosis. © PHOTOTAKE INC./ ALAMY.

osteoporosis because it results from taking certain drugs or having other diseases.

- Type 1: This type occurs in women after menopause and results from declining levels of estrogen and other sex hormones in the body.
- Type 2: This type of osteoporosis occurs in elderly men as well as elderly women because of decreased bone formation due to aging.
- Type 3: Type 3 osteoporosis is caused by long-term use of certain medications—particularly steroids and drugs given to treat epilepsy—and by such conditions as malnutrition, Klinefelter syndrome, Turner syndrome, thyroid disorders, hemophilia, Marfan syndrome, rheumatoid arthritis, lupus, and lymphoma.

#### Demographics

The National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) estimates that 10 million people in the United States have osteoporosis as of 2008, with another 34 million having low bone density, a condition called osteopenia. Osteopenia can develop into osteoporosis if it is not treated. Osteoporosis is responsible for more than 1.5 million fractures annually, including 300,000 hip fractures, 700,000 vertebral fractures, 250,000 wrist fractures, and more than 300,000 fractures in other parts of the body. The costs of treating osteoporosis and the fractures that it causes come to \$14 billion each year.

#### Osteoporosis in Men

Like anorexia, osteoporosis is usually considered a women's disease. However, men over the age of fifty have a one-in-four chance of having a bone fracture due to osteoporosis. In addition, men who take certain medications particularly cortisone and other steroid drugs have the same risk of developing osteoporosis as women who take these medications. More than two million American men have osteoporosis as of 2008; each year, 80,000 of these men will suffer a hip fracture, and one-third of them will die within a year.

One of the men who developed osteoporosis at a young age from taking steroid medications was U.S. President John F. Kennedy (1916-1963). Kennedy developed colitis, a chronic disease of the lower intestinal tract, and was treated for it in the 1930s and 1940s with steroid medications. It was not known at that time that long-term use of these drugs can damage bone. By the time Kennedy was elected to the presidency in 1960, he had three fractured vertebrae in his lower back due to osteoporosis. Medical records that were opened to historians in 2003 showed that he was routinely given seven or eight shots of procaine (the drug that dentists use to numb patients' mouths) before press conferences because the pain in his back was so intense.

Some people are at greater risk than others of developing osteoporosis. Some of these risk factors can be changed, while other factors cannot be altered. Risk factors that cannot be changed include:

- Sex. Women are at greater risk than men, particularly after the menopause.
- Race/ethnicity. Asian and Caucasian women have a higher risk of osteoporosis than African American or Hispanic women.
- Body build. Small-boned people of either sex are at greater risk of osteoporosis than people with average or heavy bones.
- Age. Both men and women have an increased risk of osteoporosis as they get older.
- Genetic factors. A tendency for bones to fracture easily appears to run in some families.

Risk factors for osteoporosis that people can change include:

- Low sex hormone levels. These can be raised in both men and women by hormone replacement therapy.
- Eating disorders, particularly anorexia.
- Depression. Emotional depression can be treated, most often with a combination of medication and psychotherapy.
- Low intake of calcium and vitamin D. People can change their eating habits and take vitamin or calcium supplements.
- Smoking and alcohol intake. People can quit smoking and drink in moderation.
- Getting the right amount of exercise. Bed rest or inadequate exercise can weaken bones, but so can too much exercise (such as marathon running).
- Medications. People who are taking medications that increase their risk of osteoporosis can ask their doctor about alternatives.

#### **Causes and Symptoms**

The basic cause of osteoporosis is that the loss of bone tissue occurs faster than the production of replacement bone. The increased rate of bone loss can be particularly critical if the person had a low or inadequate peak bone mass originally. A low peak bone mass can result from malnutrition in childhood; inadequate intake of calcium or vitamin D (necessary for the body to make use of calcium in the diet); an eating disorder in adolescence, when the body's need for calcium is at its height; or lack of exercise.

Osteoporosis can proceed for a long time without any noticeable symptoms. Some older adults simply notice that their height is shrinking. This loss of height is caused by compression of the bones in the spinal column. Sometimes the vertebrae fracture as they come closer together. This type of injury is called a compression fracture and may produce noticeable back pain.

Another common symptom of osteoporosis is a fragility fracture. Fragility fractures occur when a person falls from his or her standing position or a lower height and breaks a bone that would not break in a person with healthy bone. The most common locations of fragility fractures in people with osteoporosis are the wrists, the hips, and the vertebrae in the spine. The patient may experience the pain in various ways. Some describe it as sharp while others describe it as dull or nagging. In some cases, the pain gets worse when the patient is trying to walk or move around.

#### Diagnosis

Osteoporosis is most likely to be diagnosed following a fragility fracture. The doctor will take a careful history of the patient's risk factors, including a possible family history of easily broken bones as well as a medication history and questions about such lifestyle factors as exercise, diet, smoking, and drinking.

The doctor may order a blood test to rule out a thyroid disorder or to check the levels of sex hormones in the blood. An x ray of the affected part of the body will be taken. The definitive diagnostic test, however, is a test of bone mineral density, sometimes called a bone densitometry test. To take this test, the patient lies on an examination table while two x-ray beams of different intensities are aimed at the bones. The result is called a T-score. It is calculated by comparing the patient's bone mineral density to that of a healthy thirty-year-old of the same sex and race. A T-score of -1.0 or higher is normal. A score between -1.0 and -2.5 indicates osteopenia. A score below -2.5 indicates osteoporosis.

#### Treatment

Treatment of osteoporosis includes a combination of lifestyle changes and medications for most patients. With regard to medications, hormone replacement therapy (HRT) used to be considered the main way of treating osteoporosis in women. But newer studies show that HRT increases the risk of heart disease and cancer in some women. So other medications that work by slowing the process of bone loss or by increasing bone density over time are more widely prescribed. Some of these medications have the additional advantage of working well in men with osteoporosis and in people who must take steroid drugs for other health problems.

Recommended lifestyle changes that can reduce the rate of bone loss include regular exercise, particularly weight-bearing forms of exercise like walking, dancing, treadmill exercises, and jumping. Other measures include quitting smoking, taking supplemental vitamin D and calcium, and watching one's alcohol intake.

#### Prognosis

The prognosis for osteoporosis depends on its type and cause; the patient's age, sex, and ethnicity; the presence of other diseases or disorders; and the patient's willingness to follow the doctor's recommendations about medications and lifestyle changes.

People do not die from osteoporosis itself but from complications from bone fractures. These complications can include chronic pain, pneumonia, blood clots in the deep veins of the leg, or breathing disorders caused by the stooped posture that results from compression fractures in the spine. The death rate within the first six months after a hip fracture is 14 percent. Even patients who survive often have a greatly lowered quality of life.

#### Prevention

People cannot change such risk factors for osteoporosis as age, sex, and race, but they can eat properly, exercise regularly, and ask their doctor about vitamin D and calcium supplements. Women who have not yet gone through menopause should get at least 1,000 milligrams (mg) of

#### WORDS TO KNOW

**Compression fracture:** A fracture caused by the collapse of a vertebra in the spinal column, usually caused either by trauma or by weakening of the bone in osteoporosis.

- **Fragility fracture:** A fracture that occurs as a result of a fall from standing height or less. A person with healthy bones would not suffer a broken bone falling from a standing position.
- **Osteopenia:** The medical name for low bone mass, a condition that often precedes osteoporosis.

**Resorption:** The removal of old bone from the body.

- **T-score:** The score on a bone density test, calculated by comparing the patient's bone mineral density to that of a healthy thirty-year-old of the same sex and race.
- **Vertebra (plural, vertebrae):** One of the segments of bone that make up the spinal column.

elemental calcium and a minimum of 800 international units (IU) of vitamin D every day. Women who have completed menopause, anyone who must take steroid medications, and all men and women over 65 should aim for 1,500 mg of elemental calcium and at least 800 IU of vitamin D daily.

Older adults should also try to reduce their risk of falls whether or not they have osteoporosis. There are balance and strength exercises that older adults can practice at home. In addition, safety measures should be taken, including wearing properly fitted shoes with non-slip soles; checking one's house for loose rugs, poor lighting, and other hazards; installing grab bars in shower stalls; and keeping a cordless phone within easy reach in case of an accident.

#### **The Future**

Osteoporosis is likely to continue to be a serious health concern because of the aging of the American population. As people continue to live longer, the number of people with Type 2 (age-related) osteoporosis will increase. In addition, people who are at risk for osteoporosis because of sex, race, or a family history of weak bones may not be completely able to prevent the disease even by careful attention to diet and exercise. It is possible that more effective medications to prevent bone loss or restore bone density will be developed. **SEE ALSO** Anorexia; Klinefelter syndrome; Lupus; Lymphoma; Marfan syndrome; Rheumatoid arthritis; Turner syndrome

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# P



Genetic

Infection

Multiple

Injury

Other

Unknown

#### Also Known As Anxiety disorder

#### Cause

Genetic, biochemical, and psychological factors

#### Symptoms

Sudden feelings of doom, lightheadedness, chills, sweating, chest pain, shortness of breath

#### Duration

May last for years if untreated

### **m** Panic Disorder

#### Definition

Panic disorder is an anxiety disorder characterized by recurrent and unexpected panic attacks that are not caused by drugs, alcohol, or certain medical conditions like asthma. It can be a disabling condition, causing people to quit jobs or social activities, avoid going outside their home, or give up exercising. It also commonly leads to frequent and unnecessary visits to hospital emergency rooms.

#### Description

The most noticeable symptom of panic disorder is a panic attack, which is defined as a period of intense fear that comes on suddenly and builds to a peak within a few minutes (usually about ten minutes). The attack is often accompanied by a general feeling of doom and an urge to leave or escape from the setting in which the attack occurs. There may or may not be a pattern to a person's panic attacks. Some people may have one or two a week over a period of time, while others may have several attacks close together and then go for months or years before another attack.

Although panic attacks are a symptom of panic disorder, the two are not the same thing. It is possible for a person to have panic attacks without being diagnosed with panic disorder. To meet the official criteria for a diagnosis of panic disorder, a person must have recurrent and unexpected

#### Tips for Handling Panicky Feelings (Source: Anxiety Disorders Association of America)

Don't just stand there—move! Getting some physical exercise, whether it is yoga, jogging, or just going for a walk, can often lower one's anxiety level.

Talk to someone—a friend, parent, spouse, or doctor. Keeping anxious feelings inside often makes them worse.

Keep a diary or daily journal. It's a good way to find out what places, events, or other things stir up anxiety or panic attacks.

Eat a well-balanced diet; don't skip meals or eat a lot of junk food. It's also a good idea to cut down on coffee, tea, cola, or other drinks that contain caffeine. panic attacks followed by at least one month of worry about having another panic attack. In addition, the official criteria specify that a person must have at least four of the following thirteen symptoms that characterize panic attacks:

- Heart palpitations or fast heartbeat
- Sweating
- Trembling or shaking
- Shortness of breath
- Choking sensations
- Chest pain
- Nausea or pain in the abdomen
- Dizziness or lightheadedness
- Tingling sensations
- Chills or hot flushes
- Depersonalization (a feeling that the self is unreal) or derealization (a feeling that the external environment is unreal)
- Fear of losing control ("going crazy")
- Fear of dying

There are two subtypes of panic disorder: panic disorder with agoraphobia and panic disorder without agoraphobia. Agoraphobia is an unrealistic fear of going outside the safety of the home, particularly to use public transportation or visit large public places (shopping malls, sports arenas, or even parks or recreational facilities). About a third of Americans diagnosed with panic disorder have panic disorder with agoraphobia. Panic disorder with agoraphobia develops when the person's fear of recurrent panic attacks takes the form of avoiding places from which a quick exit or escape might be difficult or embarrassing (such as airplanes, trains, or buses; crowded stores; worship or other group activities, etc.). This form of panic disorder can interfere significantly with people's ability to complete their education, go to work, or meet friends outside the home.

#### Demographics

According to the National Institute of Mental Health (NIMH), between 1 and 4 percent of the American population suffers from panic disorder,

most of them adolescents or young adults. About 6 percent of the population will have a panic attack at some point in their lives. The average age of patients with panic disorder is twenty-four. It is unusual for people over forty-five to develop panic disorder for the first time.

Women are between two and three times more likely than men to develop panic disorder. With regard to race and ethnicity, researchers disagree. Some studies suggest that African Americans have a slightly higher rate of panic disorder than either Caucasian or Asian Americans, while other researchers think that these findings point to problems in the screening interviews used to detect panic disorder rather than the actual rates of occurrence.

People with certain medical conditions, such as hypertension (high blood pressure), migraine headaches, and irritable bowel syndrome, appear to have an increased risk of developing panic disorder.

#### **Causes and Symptoms**

The causes of panic disorder are not completely understood. Some possible causes that researchers are studying include:

- Genetic factors. A first-degree relative (parent, child, or sibling) of a person with panic disorder is eight times as likely to develop it as a person in the general population.
- Biochemical imbalances in the central nervous system. Some doctors think that differences in body chemistry cause some people to overreact to noises, lights, or other stimuli in the environment.
- Consuming large amounts of coffee, tea, or alcohol. These substances appear to trigger panic attacks in some people.
- A tendency to convert anxious thoughts into physical symptoms.
- A tendency to intensify minor physical sensations into the physical symptoms of a full-blown panic attack. For example, a man whose heartbeat speeds up when he is angry may worry when he notices the change in heart rate. He then experiences the resulting anxiety as the chest pain of a panic attack.

#### Diagnosis

The diagnosis of panic disorder is usually a diagnosis of exclusion, which means that the doctor arrives at the diagnosis by ruling out other possibilities rather than by positively identifying the disorder on the basis of tests. There are no laboratory tests or imaging studies for panic disorder. The doctor will examine the patient for such medical conditions as disorders of the thyroid gland; asthma or other breathing disorders; or substance abuse. Men in particular are likely to drink alcohol in order to cope with panic attacks. The doctor will want to make sure that the patient is not abusing alcohol or prescription drugs.

If people go to the emergency room complaining of chest pain, the doctors there will commonly ask two simple questions to screen for panic disorder: 1) Have they had a spell or attack in the past six months when all of a sudden they felt anxious, frightened, or very uneasy? 2) In the past six months, have they ever had a spell in which they felt their heart race, could not catch their breath, or felt faint? A "yes" answer to either question is considered a positive screen for panic disorder.

#### Treatment

Panic disorder is highly treatable. Most patients are given a combination of medications and psychotherapy. The two forms of psychotherapy that are most useful in treating people with panic disorder are exposure therapy (for those with agoraphobia) and cognitive behavioral therapy. In exposure therapy, patients are introduced to their feared situation in gradual stages until they feel comfortable with it. For example, someone who is afraid to go shopping for groceries might start by just opening the front door, then walking down the front path to the sidewalk. Next, the person walks a few blocks, then goes to the store itself and purchases only one item, and so on. Some people recovering from agoraphobia refer to exposure therapy as the "five Rs," which stand for react, retreat, relax, recover, and repeat.

In cognitive behavioral therapy (CBT), patients are given some education about anxiety and are taught to recognize and control their reactions to panic attacks. In CBT sessions, people learn to recognize things that trigger panic attacks or make them worse, such as specific thoughts or situations. They also learn to modify behavior so that it is more useful than simply avoiding the feared place or situation.

Doctors may prescribe medications in addition to psychotherapy, particularly if the patients' panic attacks are keeping them virtually housebound. The medications prescribed for adults are usually antidepressants or tranquilizers. It may take several weeks for the medications to take effect. The doctor may need to try more than one medication before

#### WORDS TO KNOW

**Agoraphobia:** An irrational fear of venturing outside the home or into open spaces, so strong that a large number of activities outside the home are limited or avoided altogether. Agoraphobia is often associated with panic attacks.

- **Cognitive:** Pertaining to thinking, learning, or memory.
- **Diagnosis of exclusion:** A diagnosis that the doctor arrives at by ruling out other diseases one by one rather than making the diagnosis on the

basis of laboratory tests or imaging studies, or other test results.

- **Panic attack:** An episode of intense fear that lasts for several minutes and is accompanied by physical symptoms or temporary disturbances of thinking.
- **Phobia:** An unfounded or morbid dread of a specific object or situation that arouses feelings of panic.

finding the drug that works best for the specific patient, but most patients with panic disorder are helped by these drugs.

Complementary and alternative therapies that are effective in treating panic disorder include yoga, meditation, relaxation techniques, guided imagery, and hypnosis.

#### Prognosis

Most people do very well when treated for panic disorder provided they stick with their treatment plan. In addition, people who have been treated with psychotherapy can usually make arrangements for "booster sessions" if they feel the need for further help. There are also many support groups and online communities for people with panic disorder.

#### Prevention

There is no known way to prevent panic disorder because its causes are not yet fully understood. People can, however, lower their risk of panic attacks by avoiding the use of recreational drugs and learning stress management or relaxation techniques.

#### The Future

Panic disorder is likely to continue to be a disabling condition for many people, particularly those who do not know that it is treatable. It is

possible that further research will help doctors learn more about the causes of the disorder and develop even more effective treatments.

SEE ALSO Alcoholism; Hypertension; Irritable bowel syndrome

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### Parasitic Diseases

A parasite is an organism that lives in close association with another plant or animal of a different species. The organism on which the parasite lives or feeds is called the host. Animal parasites cause harm to their hosts in many different ways, by irritating or damaging the skin or hair, robbing the host of nourishment, causing blood loss, or carrying bacteria and viruses that cause disease.

Parasitic diseases are a group of illnesses caused by organisms that live either inside the body, like protozoa (one-celled organisms) or helminths (worms); or outside the body, like lice and fleas. Some parasitic diseases that affect humans are carried by insects or other animals. These are called vectors. For example, rat fleas are vectors of plague. Infected fleas live as parasites on the rats and pass on the plague bacteria to humans when the rats die and the fleas need a new host. Similarly, cats are vectors of toxoplasmosis. This disease is caused by a protozoan (one-celled organism) that lives inside cats but can infect humans who touch cat feces containing the organism.

Parasitic diseases are a challenge to public health doctors because there are no vaccines available to prevent these illnesses. Controlling and treating these diseases depends almost entirely on medications, good sanitation, and careful attention to personal cleanliness and safe food handling practices.

**SEE ALSO** AIDS; Anemias; Lice infestation; Lyme disease; Malaria; Plague; Toxoplasmosis

### **u** Parkinson Disease

#### Definition

Parkinson disease, or PD, is an age-related movement disorder characterized by tremor (repeated rhythmic shaking or trembling); short, jerky movements and muscular rigidity; difficulty keeping one's balance; walking with a stooped posture and without the associated arm swinging; and slowing down of voluntary movements. It is named for James Parkinson (1755–1824), a British doctor who first described it in 1817.

#### Description

Parkinson disease is a disorder that affects the patient's ability to move smoothly and at a normal rate. It is a progressive disease, meaning that the symptoms get worse over time. It develops gradually over a period of years (usually about twenty years) and tends to affect only one side of the body at first. Patients may notice trembling or shaking in only one hand, which comes and goes and usually disappears during sleep. The tremor might involve the lips or chin rather than the hand. Patients might also notice a slight dragging of one foot when they walk. Other early symptoms include fatigue, depression, constipation, and sleep **Also Known As** PD, paralysis agitans, shaking palsy

#### Cause

Destruction of nerve cells in the parts of the brain that control movement

#### Symptoms

Tremor, rigidity, slowingdown of movement, loss of balance

**Duration** Years
#### Parkinson Disease

Computer image of the brain, with areas involved in Parkinson disease colored in yellow. DAVID GIFFORD / PHOTO RESEARCHERS, INC.



problems; none of these are unique to PD and are often attributed to other disorders or simply to the aging process.

The next symptoms to appear are typically increasing difficulties with gait (a person's normal pattern of walking), coordination, and keeping one's balance. Some patients notice that it takes them longer to get dressed, to write notes, or complete other tasks that require flexibility in their hands. They may find that they cannot type or play the piano without making many more errors than they used to.

Most patients with PD eventually develop depression, anxiety, memory loss, and problems with speech or swallowing as well as difficulties with movement. Although it is unusual for patients with Parkinson disease to suffer from dementia (loss of thinking and problem-solving abilities in the early stages of the disorder, some do develop dementia—including hallucinations—in its later stages.

## Demographics

Parkinson disease is almost entirely a disorder of older adults. It is one of the most common movement disorders in the elderly, affecting about 1 percent of adults over sixty in the United States. It is rare for people younger than forty to be diagnosed with PD. Worldwide, the frequency of Parkinson disease is thought to be about 120 persons in every 100,000. Men are one-and-a-half times as likely as women to develop Parkinson disease. PD appears to affect most races and ethnic groups equally. There are a few exceptions, such as the Parsis population in India, which have a higher than average rate of PD.

People who have a parent or sibling with Parkinson disease have a slightly increased risk (5 percent) of developing the disease themselves.

#### **Causes and Symptoms**

The cause or causes of Parkinson disease are not completely understood. Seven different gene mutations have been identified since the 1990s in Italian, Greek, English, and German families with a history of PD. Researchers are not yet certain, however, whether there are additional genes that play a role in the development of PD, and if so, how they interact to produce the symptoms of the disease. Another area of research is the role of pesticides in possibly triggering PD, although this theory is still debated. It is clear, however, the many people developed Parkinsonism after the influenza pandemic in 1918–1919, but the modern strains of influenza do not have this association.

What is known is that the tremor and other muscle-related symptoms of Parkinson disease are caused by damage to a part of the brain called the substantia nigra. The cells in this area produce a chemical called dopamine, which transmits nerve signals between the substantia nigra and another part of the brain that governs movement. Most Parkinson patients have lost 60 to 80 percent or more of the dopamineproducing cells in the substantia nigra by the time the first symptoms appear. The reason for the loss of these cells, however, has not yet been identified.

There are four symptoms of PD that are considered primary symptoms:

- Tremor. This is often the symptom that prompts patients to see their doctor.
- Muscle rigidity. Some patients with PD have muscles that are so stiff they cause pain.
- The slowing down of normal voluntary movements.
- Impaired balance. The patient may walk with a shuffling gait as well as being at high risk of falling. This symptom is usually the last of the primary symptoms to appear, usually about eight years after the early symptoms.

Other symptoms that may or may not occur in patients with PD include:

- The face lacks expression; may look like a mask.
- Anxiety and depression. Many patients become fearful and insecure in social situations, or irritable and moody.
- Difficulty with swallowing and chewing. The patient may drool or have difficulty eating enough food to maintain good nutrition.
- Speech problems. About 50 percent of patients with PD have difficulty talking; they may slur their speech, speak too softly, or speak too fast.
- Skin problems. The skin may become either very oily or extremely dry.
- Heavy sweating.
- Constipation.
- Difficulty sleeping. The patient may have difficulty falling asleep at night, have vivid nightmares, or may fall asleep suddenly during the day.
- Dementia and cognitive (thinking-related) problems. The dementia that occurs in PD may affect the patient's memory, language, reasoning, or other mental skills; it occurs in about 30 percent of patients.
- Fatigue.

### Diagnosis

The diagnosis of Parkinson disease can be difficult because there is no single diagnostic test for the disorder. The symptoms of PD can occur with several other disorders, including stroke, head injuries, carbon monoxide poisoning, the side effects of certain medications, and hydrocephalus. Misdiagnosis is not unusual; in the recent past, when diagnosis was based on the patient's having two of the four primary symptoms of PD, the diagnosis was incorrect in 25 percent of cases.

The diagnosis of PD is usually based on taking the patient's history, including a family history of Parkinson disease if any, and a medication history. Blood tests and imaging studies are not helpful because the results are usually normal in patients with Parkinson disease. In some cases the doctor may need to see the patient several times over a period of months to determine whether the symptoms are consistently present.

The patient may be referred to a neurologist who specializes in movement disorders for a detailed evaluation. The specialist will examine the patient's reflexes and other parts of the nervous system as well as the ability to move normally. The patient might, for example, be asked to walk across the room and back, write a note, or perform other simple tasks involving the hands. The findings that are considered the most accurate indicators of PD are:

- Two or more of the following primary symptoms: tremor, muscle rigidity, and slowed motion.
- Symptoms on only one side of the body.
- Tremor that is more noticeable when the patient's hands are resting in the lap than when they are moving.
- A strong response to levodopa, a drug that is used to treat PD.

## Treatment

To date, there is no cure for Parkinson disease. Medications are the first line of treatment for PD. The first drugs that are prescribed are drugs that give the person greater control over muscle rigidity and other movement problems, especially levodopa, a drug that is taken by mouth which the body can convert into dopamine. However, levodopa can have serious side effects, including involuntary movements, mental confusion, and hallucinations. In addition, it usually works for only four to six years before it starts to lose its effectiveness in controlling the patient's symptoms.

Other drugs that may be given include drugs like bromocriptine; they mimic the effects of dopamine on the brain rather than being converted into dopamine itself. Still other drugs may be given to inhibit the effects of an enzyme in the brain that breaks down dopamine. These drugs help to prolong the effects of levodopa and may eventually allow the doctor to reduce the person's dose of levodopa. A newer medication, selegiline, appears to slow down the destruction of the cells in the substantia nigra, and is often given to patients in early-stage PD.

In addition to drugs to help the patient control movement, the doctor may also prescribe antidepressants if the person has become depressed, or antipsychotic medications if the person is having hallucinations. The types of drugs prescribed and the dosages are individualized, since patients diagnosed with PD do not all have the same symptoms with the same severity.

There are some surgical treatments used for patients with advanced PD. One option is surgery that destroys specific parts of the brain that contribute to tremor or muscle rigidity. This type of surgery is

permanent, however, and has been largely replaced by deep brain stimulation or DBS. DBS is a procedure in which an electrode is implanted within the brain and connected to a pulse generator implanted beneath the patient's collarbone. The pulse generator stimulates the brain periodically with a mild electrical current that eases at least some of the symptoms of Parkinson disease. DBS cannot, however, be used with people with memory problems, hallucinations, a poor response to levodopa, severe depression, or general poor health.

Physical therapy, occupational therapy, speech therapy, and support groups are also recommended forms of treatment for PD.

### Prognosis

PD is not by itself a fatal disease but gets worse over time; in the later stages of the disease, PD may cause complications like choking, pneumonia, and falls that can lead to the patient's death. The speed of progression varies from patient to patient; some patients may be able to function relatively well for nearly twenty years after diagnosis while others become completely dependent on caregivers within a few years. There is no way to predict how quickly the disease will progress in an individual patient. Patients who are diagnosed with Parkinson disease are usually advised to plan ahead for the time when they may require a fulltime caregiver or transfer to a nursing home.

### Prevention

There is no known way to prevent Parkinson disease because its causes are still being investigated.

## The Future

One area of investigation in Parkinson disease is the search for a biomarker (abnormal body chemical) in patients with PD that could be used as a screening tool to identify people at increased risk of developing the disease before the first symptoms appear. Another area of research is the use of an imaging technique called positron emission tomography (PET). PET may allow scientists to understand more about the disease process and the possible causes for the loss of the nerve cells that produce dopamine. Still a third field of investigation is the hunt for additional genes and gene mutations involved in PD.

## WORDS TO KNOW

**Biomarker:** A substance produced by the body that is distinctive to a particular disease and can be used to identify its presence or track its progress.

**Dementia:** Loss of memory and other mental functions related to thinking or problemsolving. **Dopamine:** A chemical produced in the brain that is needed to produce smooth and controlled voluntary movements.

Gait: A person's characteristic pattern of walking.

**Tremor:** An unintentional shaking or trembling movement. Tremor is a characteristic symptom of PD.

SEE ALSO Alzheimer disease; Depression; Hydrocephalus; Stroke

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# Patau Syndrome

# Definition

Patau syndrome is a genetic disorder caused by the presence of an extra copy of chromosome 13 or by a portion of chromosome 13 translocated (attached) to another chromosome plus two copies of chromosome 13.

# Description

Babies born with Patau syndrome have a characteristic set of facial, neurological, and heart defects and a very high mortality rate.

# Demographics

Estimates of the frequency of Patau syndrome range from one in every 5,000 live births to one in every 12,000. The syndrome occurs with equal frequency in all races and countries. The chief risk factor for Patau syndrome is the age of the mother; the average age of mothers of babies with the syndrome is thirty-one.

It is thought that male and female fetuses are equally likely to be affected at the time of conception; however, the mortality rate among boys is higher at all points along the baby's development before and after birth.

# **Causes and Symptoms**

Patau syndrome can result from genetic errors during the formation of germ cells (eggs and sperm) or during cell division shortly after the egg is fertilized by the sperm. The most common form of Patau syndrome, which accounts for about 75 percent of cases, occurs when an egg or sperm carrying two copies of chromosome 13 is involved in conception. The reason for the extra copy in the abnormal germ cell is a genetic error called nondisjunction. During the normal process of germ cell formation, the paired chromosomes in the cell divide so that each daughter cell has only one member of the pair. In nondisjunction, one daughter cell gets both members of the chromosome pair and the other cell has none. If a germ cell carrying two copies of chromosome 13 is fertilized by a normal

### Also Known As

Trisomy 13, Bartholin-Patau syndrome, D1 trisomy

#### Cause

An extra copy of human chromosome 13

#### Symptoms

Severe mental retardation, brain malformations, extra fingers or toes, heart defects, abnormal genitals, death shortly after birth

#### Duration

Lifelong

#### Patau Syndrome

Patient's hand deformed by Patau syndrome. BIOPHOTO ASSOCIATES / PHOTO RESEARCHERS, INC.



germ cell from the other parent, the child will have three copies of chromosome 13. This genetic error is called a full trisomy 13.

About 20 percent of cases of Patau syndrome develop when a part of chromosome 13 becomes attached to chromosome 14 either before or at the moment of conception. This type of genetic error is called a translocation. The child will be born with two copies of chromosome 13 plus some extra genetic material from chromosome 13 attached to chromosome 14. A child with this type of genetic error is said to have partial trisomy 13.

About 5 percent of cases of Patau syndrome occur in children who have some body cells with the extra copy of chromosome 13 and some body cells without the extra copy. This condition is called mosaic Patau. It is thought to result from random errors in cell division during the early stages of fetal development. Children with mosaic Patau are usually less severely affected than those with full or partial trisomy 13 and may lack many of the features associated with Patau syndrome.

The symptoms of Patau syndrome include a high mortality rate even before birth. Many embryos with full trisomy 13 die during pregnancy or are expelled from the mother's womb in what is called a spontaneous abortion or miscarriage. Those who survive until birth usually live only a few days; 69 percent die of breathing problems and another 13 percent die during this period from heart defects. Although some children with

# **Rasmus Batholin and Klaus Patau**

The two doctors who are responsible for identifying Patau syndrome are Rasmus Bartholin (1625–1698), a Danish doctor, and Klaus Patau (1908–1975), an American geneticist who was born in Germany. Bartholin was a professor of medicine at the University of Copenhagen who wrote the first description of a baby with Patau syndrome in 1657. His discovery is the reason why the syndrome is sometimes called Bartholin-Patau syndrome.

Doctors in the seventeenth century did not have the research tools that allow modern doctors to study the way human cells reproduce, and so Bartholin could not identify the syndrome he described as a genetic disorder. It was not until the late 1950s when Patau, a researcher in the Department of Genetics at the University of Wisconsin-Madison, was able to identify the extra chromosome 13 as the cause of the syndrome that was later named for him. Patau published the first article about Patau syndrome in a British medical journal in 1960. Patau syndrome do not have all the physical features that characterize the syndrome, the following are considered typical:

- Holoprosencephaly. This term refers to the failure of the infant's brain to divide into two equal halves during its development before birth. Babies with holoprosencephaly usually have facial abnormalities, including cleft lip, cleft palate, a misshapen or completely absent nose, and abnormally small eyes placed unusually close together. The entire head may also be unusually small.
- Heart defects. Eighty percent of infants with Patau syndrome are born with one or more heart defects, including an opening between the two lower chambers of the heart or the heart being situated on the right rather than the left side of the body.
- Polydactyly. This term refers to the presence of extra fingers or toes.
- Spina bifida. Spina bifida is a condition in which the spinal cord is partially open at birth instead of being covered by the bones of the spinal column.
- Abnormal genitalia.
- Rocker-bottom feet. This term is used to describe abnormally long and slender feet with pointed heels turned outward like the bottom rails of a rocker.
- Low-set ears and abnormalities of the inner ear.
- Abnormal kidneys.

## Diagnosis

Diagnosis of Patau syndrome is usually made on the basis of the child's appearance at birth. It can be diagnosed before birth on the basis of ultrasound studies during the first three months of pregnancy; a sample of the

mother's blood plasma; or by genetic analysis of cells taken from the baby's blood or the amniotic fluid that surrounds the baby inside the womb.

### Treatment

There is no treatment for Patau syndrome itself. Infants who survive the first two days often require two or more weeks of treatment in an intensive care unit (ICU). Those who survive the first six months may be treated with surgery for specific heart defects and other abnormalities; however, their mental retardation cannot be corrected by surgery and they remain at high risk of developing severe curvature of the spine in adolescence. They also have an increased risk of developing cancer.

## Prognosis

The prognosis for children with Patau syndrome is very poor. Of those who do not die before birth, the average length of survival is 2.5 days. Only one child in twenty lives longer than six months. A few children live into their teens, and there are a few case reports of people living into their early twenties with Patau syndrome. Most long-term survivors are female.

## Prevention

Since Patau syndrome is thought to be caused by a spontaneous genetic mutation rather than an inherited genetic defect, there is no way to prevent it. Pregnant women over thirty-five should have tests during the first trimester (three-month period) of pregnancy to screen for the syndrome. These tests may involve ultrasound studies, which can detect abnormalities in the baby's heart or facial development, followed by a photographic analysis of cells taken from the fluid that surrounds the baby in the womb. This analysis, or karyotype, is needed to distinguish Patau syndrome from other genetic disorders that can cause heart defects and facial abnormalities. Doctors recommend that the parents of a child with Patau syndrome should consult a genetic counselor for advice about future pregnancies.

## **The Future**

It is not known whether the increasing numbers of women having children in their thirties or forties will lead to an increase in the number of

# WORDS TO KNOW

- **Germ cell:** A cell involved in reproduction. In humans the germ cells are the sperm (male) and egg (female). Unlike other cells in the body, germ cells contain only half the standard number of chromosomes.
- **Holoprosencephaly:** A disorder in which a baby's forebrain does not develop normally. The infant's brain fails to divide into two cerebral hemispheres; this failure in turn leads to facial deformities and abnormal brain structure and function.
- **Karyotype:** A photomicrograph of the chromosomes in a single human cell. Making a karyotype is one way to test for genetic disorders.
- **Mosaicism:** A condition in which a person has some body cells containing an abnormal number of chromosomes and other cells containing the normal number. Mosaicism results

from random errors during the process of cell division that follows conception.

- **Nondisjunction:** A genetic error in which one or more pairs of chromosomes fail to separate during the formation of germ cells, with the result that both chromosomes are carried to one daughter cell and none to the other. If an egg or sperm with a paired set of chromosomes is involved in the conception of a child, the child will have three chromosomes in its genetic makeup, two from one parent and one from the other.
- **Translocation:** A genetic error in which a part of one chromosome becomes attached to another chromosome during cell division.
- **Trisomy:** A type of genetic disorder in which a cell contains three copies of a particular chromosome instead of the normal two. Patau syndrome is one of several trisomies.

children born with Patau syndrome. Many women whose fetuses are diagnosed with Patau syndrome choose to end their pregnancies before childbirth.

There are online support groups for families of children with trisomy 13; one posts photo albums of long-term survivors of Patau syndrome as well as information about the disorder and frequent updates about medical research.

SEE ALSO Down syndrome; Edwards syndrome

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# Peptic Ulcers

See Ulcers.



# Periodontal Disease

## Definition

Periodontal disease refers to inflammation of the gums and damage to the tissues beneath the gums caused by bacteria that live in the mouth. Gingivitis is the term that dentists use for inflammation that is limited to the gums. Disease that affects the tissues and bone beneath the gums is called periodontitis. In severe cases, periodontal disease can lead to the loss of teeth. There is also some possibility that it can also lead to an increased risk of heart disease and stroke, but researchers are still studying the issue.

## Description

Periodontal disease is a disorder of the gums that can lead to sore, swollen gums that bleed easily or even to the eventual loss of teeth. It develops slowly over a period of years in most people and often has no early symptoms at all. The first stage of periodontal disease is called gingivitis or inflammation of the gums. The inflammation is caused by bacteria that live in the mouth in a sticky film called plaque. The bacteria can attack the teeth along the gum line if they get into the small space between a tooth and the gum known as a sulcus. If a person does not remove the plaque by regular brushing and flossing, the bacteria in or near the sulci Also Known As Gum disease, gingivitis

#### Cause

Bacterial infection of the gums

#### Symptoms

Gums that bleed easily, swollen or sore gums, painful chewing, tooth loss, bad taste in mouth

**Duration** Years

#### Periodontal Disease

X ray of periodontal disease. The second tooth from the left has a large abscess at its root, and three teeth show bone loss. © PHOTOTAKE INC. / ALAMY.



release toxins that damage the gums and cause them to pull away from the base of the teeth. The person may notice that his or her gums look red or swollen, or that the gums bleed after brushing or biting into a hard food like an apple. The person may also have a bad taste in the mouth.

The next stage in periodontal disease is called periodontitis. If the gingivitis is not treated, the bacteria in the plaque multiply inside the pockets that are formed as the sulcus at the base of each tooth is enlarged. The bacteria continue to secrete toxins, and the body's immune response to the infection combines with the toxins to break down the connective tissues underneath the gums and the underlying bone. If this process is not stopped by appropriate treatment, the teeth become loosened and may fall out or have to be pulled.

## Demographics

Periodontal disease is a common health problem among adolescents and adults in developed countries. It is estimated that 50 percent of teenagers and 80 percent of adults in the United States have some form of gum disease. Periodontal disease—not aging—is the largest single cause of tooth loss.

There are a number of risk factors for periodontal disease:

• Smoking or chewing tobacco. Tobacco use is the largest single risk factor for gum disease because it lowers the immune system's

response to the bacteria that live in the mouth. It also interferes with the effectiveness of treatments for periodontal disease.

- Diabetes. Poor control of blood sugar levels speeds up the loss of soft tissue and bone in the mouth.
- Medications. Birth control pills, anti-seizure drugs, steroid medications, and anticancer drugs can all increase the risk of gum disease.
- False teeth that don't fit properly.
- Crooked teeth.
- Poor nutrition.
- Pregnancy. The hormonal changes of pregnancy make a woman's gums more vulnerable to infection.
- Emotional stress. High stress levels weaken the body's immune system, including its ability to fight off bacteria in the mouth.
- Fillings that have cracked or become loose.
- Genetic factors. In the United States, African Americans have higher rates of periodontal disease than members of other ethnic or racial groups. In Europe, people from the countries closer to the Mediterranean Sea have higher rates of periodontal disease than people from northern Europe.

# **Causes and Symptoms**

The basic cause of periodontal disease is infection of the gums by bacteria living in the plaque that forms on the teeth and the gums after a person eats. The intensity of the body's immune response to the inflammation which varies from person to person—is an additional factor, as are diseases like diabetes or AIDS that affect the whole body.

The symptoms of periodontal disease include:

- Sore or swollen gums
- Bleeding from the gums after brushing or flossing
- Bad breath
- An unpleasant or metallic taste in the mouth
- Visible shrinking of the gums and exposure of the roots of the teeth
- Increased sensitivity of the teeth to temperature changes
- Teeth that feel loose in the mouth
- A change in the way the teeth fit together when the person bites

## Diagnosis

The diagnosis of periodontal disease can often be made when the dentist simply looks inside the person's mouth. The dentist may also use a probe to measure the size of the pockets between the teeth and the gums, and will usually take x-ray films of the teeth to see whether the connective tissues and bone beneath the teeth have been damaged.

# Treatment

Gingivitis can be treated during a routine dental checkup, although the treatment may take more than one appointment. A dental hygienist can assist the dentist in removing plaque and tartar from the area around the gum line by scaling and planing the teeth. Scaling involves the use of a scraper to remove plaque from above and below the gum line. Planing involves smoothing out the roots of the teeth to encourage the gums to reattach to the roots.

Other treatments that may be needed to treat periodontal disease include:

- Antibiotic mouthwash to kill the bacteria that live in plaque. The dentist may prescribe a mouthwash containing a drug called chlorhexidine. The mouthwash is used at home like a regular mouthwash.
- Antibiotic chips or gels. These are placed in the gum pockets after scaling and planing. The antibiotic is slowly released over a period of about a week.
- Enzyme suppressant. This is a low-dose oral medication to be taken at home after scaling and planing. It is given to lower the body's immune response to the bacteria in the mouth.
- Surgery. If the gum disease has progressed to the point where tissue has been lost, the dentist may refer the patient to a specialized dentist called a periodontist. The periodontist may pull back the gums to remove plaque and tartar that cannot be removed by planing and scaling, and then suture (sew) the gums back in place around the base of the teeth. Another type of surgery that is sometimes used involves bone and tissue grafting to prevent the loss of teeth.

## Prognosis

The prognosis of periodontal disease depends on the stage at which it is diagnosed and treated. Gingivitis can almost always be treated without

# WORDS TO KNOW

- **Gingivitis:** The medical term for inflammation of the gums.
- **Periodontitis:** The medical term for gum disease that involves the connective tissue and bone beneath the gums.
- **Plaque:** A film that forms on the surface of the teeth and gums containing bacteria, saliva, and dead cells.

**Scaling:** Scraping tartar away from the teeth around the gum line.

**Sulcus (plural, sulci):** The space or crevice between a tooth and the surrounding gum tissue.

Tartar: Hardened plaque.

loss of teeth. The prognosis of periodontitis depends on the individual's overall health and the extent to which the infection has damaged the tissues and bone underneath the teeth. Diabetics, patients with AIDS, and heavy smokers generally have a poorer prognosis.

## Prevention

Periodontal disease is preventable with proper care of the mouth. The American Dental Association (ADA) recommends the following preventive steps:

- Get regular dental checkups that include professional gum cleaning if needed.
- Brush and floss after eating, and at least twice a day for a period of about three minutes.
- Get a fresh toothbrush every three months.
- Use a brush with soft rather than hard bristles; hard bristles can irritate the gums and make it easier for the bacteria to invade the injured tissues.
- Use a toothpaste that contains fluoride. The dentist may also recommend a mouthwash that contains fluoride.
- Avoid eating too many sweets and other foods that encourage plaque to form on the teeth and gums.
- Quit smoking (or do not start).

### The Future

Some recent studies have suggested that periodontal disease increases a person's risk of stroke or heart disease in later life or a woman's risk of delivering a baby prematurely. Other researchers think there may be a connection between periodontal disease and increased difficulty in controlling blood sugar levels in people with diabetes. Although researchers have not proved a cause-and-effect relationship between gum disease and these other disorders, they have provided a reminder that proper care of the mouth and teeth is as important to good health as care of other body systems.

SEE ALSO Diabetes; Smoking; Stress; Tooth decay

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# Phenylketonuria

# Definition

Phenylketonuria, or PKU, is an inherited disease caused by a mutation in a gene called the PAH gene on chromosome 12. This gene affects the body's ability to use phenylalanine, one of the amino acids that are the building blocks of proteins.

# Description

Children with PKU cannot use phenylalanine, an amino acid that occurs in all protein foods and some artificial sweeteners, particularly aspartame. As a result, the amino acid and two closely related substances build up to harmful levels in the child's body, causing mental retardation and other serious health problems.

Not all children with PKU are as severely affected as those with the so-called classic form of the disorder. Children with classic PKU look normal until they are a few months old, although they may appear to have fairer skin and lighter hair than their parents and siblings. They may also suffer from eczema and other skin disorders, or have a musty or mouse-like odor on their breath or in their sweat or urine that results from the high levels of phenylalanine in the body.

The severe symptoms of classic PKU begin several months after birth, when the baby begins to have seizures, its head appears abnormally small in proportion to the rest of the body, and normal mental development slows down. In later life such children may become hyperactive or develop other neurological problems.

Babies who are born to mothers with uncontrolled PKU (mothers who no longer follow the special PKU diet) are at high risk of being born with mental retardation because they are exposed to high levels of phenylalanine in the mother's blood before birth. These infants may also have a low birth weight and grow more slowly than other children. In addition, the mother has an increased risk of miscarriage before the baby is born.

# Demographics

Phenylketonuria is more common in some countries and ethnic groups than in others. In the United States, it affects about one child in every Also Known As PKU, Følling's disease

#### Cause

Mutation in the PAH gene on chromosome 12

#### Symptoms

Small head, mental retardation, heart problems, seizures, developmental delays

**Duration** Lifelong



A newborn being tested for PKU by a heel prick. © PICTURE PARTNERS / ALAMY.

10,000–15,000; however, in Turkey it affects one child in every 2,600. High rates of PKU also occur in Ireland, certain parts of Yemen, Eastern Europe, Italy, and China. The country with the lowest rate of PKU is Finland, where only one child in 100,000 is diagnosed with PKU.

In the United States, PKU is more common among Caucasians and Asians than in other races and ethnic groups. It affects males and females equally.

## **Causes and Symptoms**

PKU is caused by a mutation in the PAH gene on chromosome 12. As of 2008 more than

400 mutations in this gene have been identified that can cause PKU. A child must inherit defective genes from both parents in order to develop PKU. Children who have not inherited defective genes from both parents, however, can still be carriers of a defective gene from one of their parents. If such a carrier marries another carrier, the couple has one chance in four of having a baby with PKU.

A person with a defective PAH gene from both parents will lack an enzyme that is needed to convert phenylalanine to other compounds that the body can use. As the unconverted phenylalanine builds up in the baby's body, it begins to damage the brain and central nervous system, leading to the characteristic symptoms of PKU.

In addition to mental retardation, seizures, and the characteristic musty odor associated with PKU, children with the disorder may have the following symptoms:

- · Delayed social skills
- Hyperactivity
- · Jerking movements of the arms and legs
- Skin rashes
- Tremor
- Self-mutilation
- Tendency to hold the hands in unusual positions

# Diagnosis

Most children in the United States and other developed countries undergo mandatory screening programs shortly after birth. The test most commonly used to check for PKU in the United States is the Guthrie test, introduced in 1962, which consists of taking a blood sample from a newborn by the heelstick method. The test is sensitive enough to detect high levels of phenylalanine shortly after birth. In a few cases children may have to have a second blood test at two weeks of age to confirm the diagnosis. It is important to detect PKU as quickly as possible so that treatment can be started promptly, as children who are not treated will become mentally retarded by the end of their first year.

## Treatment

The central form of treatment for phenylketonuria is strict adherence to a diet that is low in phenylalanine. PKU is one of a small number of genetic disorders that can be treated by diet alone. The PKU diet severely restricts or eliminates foods high in phenylalanine, including breast milk, meat, chicken, fish, nuts, cheese, legumes and other dairy products. The person's intake of potatoes, bread, pizza, corn, pasta, and other starchy foods must be carefully monitored. Many diet foods and soft drinks that contain aspartame (NutraSweet) must also be avoided. In many cases the PKU diet must be supplemented with iron and other nutrients that are usually found in foods high in phenylalanine. To provide some variety at mealtimes, there are companies that make low-protein pastas, breads, imitation cheese, baking mixes, and other foods for people with PKU. These foods are covered by medical benefits in some states.

# The Doctor Who Discovered PKU

Ivar Asbjørn Følling (1888–1973) was a Norwegian physician and biochemist who identified phenylketonuria and determined its cause. His discovery began in January 1934 when a young mother consulted him about her two children, a son and a daughter, who had been normal at birth but were slowly becoming mentally retarded. When the son was about a year old, Mrs. Egeland noticed that his urine had a strange smell. She consulted Følling because she knew he was an expert in nutritional biochemistry.

Dr. Følling studied the child's urine in the laboratory and discovered that it contained a chemical related to the amino acid phenylalanine that had never before been identified in human urine. Følling then found that Mrs. Egeland's children lacked the ability to make use of phenylalanine in the diet. He then studied children in a nearby institution for the mentally retarded and found that a significant number of them had the same condition as the Egeland children. Although it took some years afterward for Følling to work out a diet that would protect children with PKU from retardation, his work has saved thousands of children since the 1930s.

Dr. Følling has been termed the most important medical scientist who failed to receive a Nobel Prize for his work. His identification of PKU was a watershed in medicine because it was the first time that a medical researcher demonstrated that a mental disorder like retardation can have a completely biochemical cause.

There is a special infant formula called Lofenalac for babies with PKU. Lofenalac can be used throughout life as a protein source that is

low in phenylalanine and balanced for the remaining essential amino acids.

At one time it was thought that teenagers over the age of eighteen and adults with PKU could stop using the special diet. As of 2008, however, doctors recommended remaining on the PKU diet for life. One reason is maintaining children's normal mental development; children who go off the special diet may lose ten points in IQ or even more. Another reason is lowering the risk of severe PKU in children born to mothers with the disease. Pregnant women with PKU who go off the special diet risk having children who are severely mentally retarded.

In 2007 the Food and Drug Administration (FDA) approved the use of Kuvan, a medication taken by mouth, which supplements the body's production of the enzyme that is needed to digest phenylalanine. The drug was considered an orphan drug until 2004, when the FDA encouraged its further development to treat PKU. Kuvan does not work for all children, however, and some doctors are concerned that children who do benefit from it (about 50 percent of children with classic PKU) may become careless about eating foods containing phenylalanine.

Some children with PKU require psychotherapy in later childhood or adolescence because of problems with self-esteem or frustration with having to follow a restricted diet.

### Prognosis

The prognosis of children (and adults) with phenylketonuria depends largely on their willingness to adhere to the PKU diet. Some children and adolescents "cheat" on the diet by eating such foods as French fries, pizza with cheese, potato chips, and bread. Vomiting, severe behavioral disorders, and loss of memory or the ability to concentrate have all been found in people with PKU who go off the special diet. Children and adults who are careful about what they eat and see a nutritionist regularly (once a month is recommended) usually do very well.

## Prevention

Women diagnosed with PKU can help their unborn children by careful adherence to the special diet. People who do not have PKU themselves but have relatives with the disorder may be carriers of the disease. They can have a genetic test before having children to determine whether they are at risk of having a child with PKU.

## WORDS TO KNOW

- **Amino acids:** A group of twenty compounds that are the building blocks of proteins in humans and other animals.
- **Heelstick:** A method for taking a sample of blood from a newborn by pricking the baby's heel with a needle and collecting a drop or two of blood on special filter paper.
- **Orphan drug:** A drug defined by the Food and Drug Administration (FDA) as intended to treat

a disease or condition that affects less than 200,000 people in the United States, or a disease of condition that affects more than 200,000 people and there is no reasonable expectation that the company can recover the costs of developing the drug.

**Phenylalanine:** The amino acid that cannot be used by the bodies of people with phenylketonuria.

# **The Future**

The FDA is conducting ongoing tests of Kuvan to see whether it could be modified to benefit more children with PKU. It is possible that other drugs will be developed to treat children with PKU who are not presently helped by Kuvan.

SEE ALSO Developmental disability; Eczema

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# Pink Eye

See Conjunctivitis.



# Plague

# Definition

Plague is an infectious disease caused by a rod-shaped bacterium called *Yersinia pestis*, which usually is spread to humans through the bites of infected fleas. Most epidemics of plague among humans have involved fleas that live in the fur of rats and other rodents. The rats are called vectors of the disease because they carry the infected fleas to other animals, including humans.

# Description

There are three major forms of plague infection in humans: bubonic, septicemic, and pneumonic. Bubonic plague is the most familiar form; it is characterized by high fever and the appearances of buboes, or tender, swollen lymph glands, in the groin, armpit, and neck of the infected person. Septicemic plague is an infection of the bloodstream; the patient may not develop buboes but has a high fever. It has a higher mortality rate than the bubonic form of plague. Pneumonic plague, the most deadly form, can be spread directly from infected humans, or sometimes cats, to other people through coughing. The plague bacteria infect the lungs, causing chest pain, coughing up of blood, and difficulty breathing along with high fever and chills.

#### Also Known As

Bubonic plague, septicemic plague, pneumonic plague

#### Cause

Bacteria transmitted to humans through infected flea bites

#### Symptoms

High fever; swollen, tender lymph glands; chills; muscle pain; severe headache; seizures

#### Duration

Possible death within a few days

#### Plague



*Blood cells with plague bacteria* (*small darks spots*). SCIENCE SOURCE.

## Demographics

The last epidemic of plague in the United States that was carried by rats occurred in Los Angeles in 1924. All cases of plague reported in North America since then have been isolated incidents. According to the Centers for Disease Control and Prevention (CDC), there are on average ten cases of plague in the United States each year, almost entirely in the Southwest and California, with the local ground squirrel as the rodent reservoir. This is a decrease from the average of eighteen cases per year in the 1980s. Worldwide, there are about 2,000 cases of death from plague every year, mostly in Africa, Asia, or South America.

Although most cases reported each year occur in Caucasians, Native Americans in parts of Utah, New Mexico, and Arizona have a high risk of getting plague because the disease is endemic in those areas and many members of these tribes work as sheepherders or ranchers.

Risk factors for getting plague include having an occupation that requires working outdoors in an area where plague is endemic or hiking and camping in such areas.

## **Causes and Symptoms**

Plague is caused by bacteria transmitted to humans, mostly through the bites of infected fleas. The fleas live in the fur of house rats, ship rats, and other rodents, and pick up the bacteria from biting an infected rodent.

# The Black Death

The Black Death is the name given to the pandemic (worldwide epidemic) of bubonic plague that began in China and moved across Central Asia to Europe in 1347. It is estimated to have killed 75 million people worldwide; between 25 and 50 million deaths occurred in Europe, representing 30–60 percent of the population at that time.

The Black Death, in part, spread to Europe through an early form of bioterrorism. The soldiers of a Mongol army besieging Caffa, a trading city on the Crimean peninsula, began to die from plague in early 1347. The Mongol general ordered the infected bodies to be catapulted over the city walls in order to spread the plague inside the city. Some Italian traders in Caffa fled back to Italy, taking the plague bacteria with them to Sicily and southern Europe. It spread northward from Italy, reaching France and Spain in 1348, England in 1349, and the Scandinavian countries in 1350.

This was neither the first nor the last pandemic of bubonic plague. The first major known outbreak was the Plague of Justinian, which began in Africa and killed about a quarter of the population of the eastern Mediterranean during the reign of the



During plague outbreaks around 1790, doctors would wear long clothing, masks, and gloves to ward off the disease. © MEDICAL-ON-LINE / ALAMY.

Emperor Justinian in 541–542 CE. The Great Plague of London later killed a fifth of the city's population between 1665 and 1666.

The fleas can then infect other rats, prairie dogs, chipmunks, rabbits, rock squirrels, and similar animals by biting them. From time to time the infected rodents begin to die off at a high rate. The fleas then go looking for other sources of blood meals, and may turn to domestic animals or humans.

When a flea sucks blood from an animal infected with plague, the plague bacteria can multiply inside the flea's upper digestive tract and cause obstruction. When the flea bites its next victim to try to feed, it injects material loaded with bacteria into the skin. Once inside the new host, the bacteria travel to the lymph nodes, where they multiply further and produce the characteristic buboes of the bubonic form of the disease. The tissue in the lymph nodes may break down and allow the bacteria to travel through the bloodstream, producing the septicemic and pneumonic forms of plague.

The incubation period of plague in humans is two to six days for the bubonic form and as little as two days for the pneumonic form. The symptoms of each form typically include:

- Bubonic plague: Rash or blister at the site of the flea bite; muscle aches; buboes; and high fever. The buboes are extremely painful; the patient may not want to let the doctor examine them and may hold the limbs in unusual positions to relieve pressure on the buboes. As the swollen lymph glands grow in size, they may burst and release blood and bad-smelling pus.
- Septicemic plague: Patients may die quickly from septicemic plague. Symptoms include sudden drop in blood pressure, nausea, vomiting, diarrhea, fever, abdominal pain, and sudden organ failure.
- Pneumonic plague: Severe cough, chest pain, difficulty breathing, and frothy or bloody material brought up by coughing.

# Diagnosis

Diagnosis is made by a combination of history-taking and recording of the patient's fever and other symptoms. A history of exposure to rodents or fleas, the presence of flea bites on the patient's skin, and a high fever are major warning signs. Because plague should be treated as swiftly as possible—within twenty-four hours—the doctor will have the patient hospitalized for antibiotic treatment. A sample of fluid from a bubo (if one is present) or cerebrospinal fluid can be sent to the CDC laboratories for analysis; most hospital laboratories are not equipped to do the specialized testing required. If the patient appears to have septicemic plague, a blood sample will be taken as well. For pneumonic plague, a sample of the material coughed up by the patient will be sent for analysis.

## Treatment

Treatment requires immediate hospitalization in an isolation room. Antibiotics are given directly into the veins or into the muscles for at least ten days. Patients dehydrated by fever will be given intravenous fluids as well. The patient may be put on a ventilator if they need help in breathing or given pure oxygen to breathe.

People who have been in contact with a patient diagnosed with pneumonic plague are often given antibiotics as a preventive measure and monitored carefully by their doctors.

Doctors are required by law to report a case of plague to the World Health Organization (WHO) as well as the CDC.

## Prognosis

Prognosis depends partly on the patient's age, general health, and the promptness of treatment. Elderly people are at high risk of dying from plague even with prompt treatment. The mortality rate of untreated patients with the bubonic form of plague is about 50 percent if sepstemic plague ensues; however, it drops to 5 percent for those who are diagnosed and treated promptly. Almost all people with pneumonic plague will die without treatment.

## Prevention

Plague can be prevented by strict control of rats in urban areas, extermination of rats on ships and in harbor areas, and by surveillance (monitoring) of prairie dogs and other rodents in areas where the plague is endemic. People who notice the sudden appearance of large numbers of dead or dying rats, mice, or other rodents in their area should notify their public health department at once, as the rapid death of these animals is a warning sign of plague.

There is no commercially available vaccine against plague.

# **The Future**

The most worrisome aspect of plague is not the possibility of an epidemic spread by fleas but rather one spread by terrorists. The CDC has identified the plague bacteria as an organism that could be turned into an aerosol form and then spread over large populations. Because people would breathe in the aerosolized bacteria, they would develop the pneumonic form of plague—the deadliest form and the form that can spread from person to person. WHO has estimated that spraying plague bacteria over a city of 5 million people would cause 150,000 cases of plague and 40,000 deaths.

## WORDS TO KNOW

- **Bioterrorism:** The use of disease agents to frighten or attack civilians.
- **Bubo:** A swollen lymph node in the neck, armpit, or groin area.
- **Endemic:** A term applied to a disease that maintains itself in a particular area without reinforcement from outside sources of infection.
- **Host:** An organism that is infected by a virus, bacterium, or parasite. Rats and other rodents are common hosts of plague.

- **Pandemic:** A disease epidemic that spreads over a wide geographical area and affects a large proportion of the population.
- **Surveillance:** Monitoring of infectious diseases by public health doctors.
- **Vector:** An animal that carries a disease from one host to another.

In order to protect against this threat, the United States, Canada, and the United Kingdom signed an agreement in April 2005 to jointly develop a more effective vaccine against plague. Two experimental vaccines, one developed in the United Kingdom and the other by the United States Department of Defense, are presently undergoing clinical trials. The vaccines will be evaluated and the one that shows greater promise will selected for advanced development.

#### SEE ALSO Anthrax

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# Pneumonia

### Definition

Pneumonia is a serious inflammatory lung disorder in which the alveoli—tiny air-filled sacs in the lungs that ordinarily absorb oxygen from the air—fill with fluid or pus. As a result of this inflammation, the person cannot get enough oxygen into the bloodstream to meet the needs of body tissues. In addition, the disease organisms responsible for most cases of pneumonia can spread from the lungs into the bloodstream and infect other vital organs, thus causing the person's death.

Pneumonia has at least thirty different causes. Although most cases of pneumonia are caused by bacteria, viruses, or other disease organisms, the illness can also be the result of non-infectious causes such as chemical injuries to the lungs, food or saliva accidentally getting into the airway, or even by allergies to substances in the air.

Also Known As Bronchopneumonia

#### Cause

Bacteria, viruses, fungi, chemicals

#### Symptoms

Fever, chest pain, shortness of breath, bloody sputum, coughing with green or yellowish mucus

#### Duration

Two to six weeks, possibly longer for the elderly

Pneumonia is sometimes classified by the ways in which people can acquire it. Some common types of pneumonia include:

- Nosocomial pneumonia: Nosocomial refers to infections acquired by hospital patients. This type of pneumonia can be very serious, partly because hospitalized people are already ill or weak from surgery, and partly because the bacteria found in hospitals are often more resistant to antibiotics.
- Ventilator-associated pneumonia: Patients who must be placed on a ventilator to

help them breathe are at increased risk of developing pneumonia.

- Community-acquired pneumonia (CAP): CAP refers to pneumonia caused by disease organisms in people who have not been recently hospitalized.
- Aspiration pneumonia: Aspiration is the medical term for getting saliva, food particles, or liquids into the airway and lungs. Aspiration pneumonia is common in the elderly, patients with Parkinson disease, and others who may have trouble swallowing normally. The materials that get into the lungs can cause pneumonia either by chemical irritation or by carrying bacteria that cause inflammation.
- Opportunistic pneumonia: Opportunistic infections are caused in people with weakened immune systems by organisms that do not ordinarily cause disease in people with healthy immune systems. People with AIDS are vulnerable to a particular type of pneumonia called PCP, which stands for *Pneumocystis carinii* pneumonia. *Pneumocystis* is classified as a fungus.
- Chemical pneumonia. Breathing in certain chemicals, particularly pesticides, can cause pneumonia.
- Walking pneumonia: Walking pneumonia is an older term for pneumonia that does not make patients sick enough to stay in bed; that is, they are well enough to go about their ordinary activities in spite of coughing and headaches. Most cases of walking pneumonia are caused by mycoplasmas, a kind of bacteria, or by viruses.

Chest x ray of pneumonia. The lower right lung (left side of the x ray) is filled with fluid. © PHOTOTAKE INC. / ALAMY.

# An Ancient Killer

Pneumonia has caused the death of many famous people, not just those who lived before the discovery of antibiotics, but also some who died in the late twentieth or early twenty-first century:

- William Henry Harrison (1773–1841), ninth president of the United States. Died one month after catching a cold at his inauguration.
- Stonewall Jackson (1824–1863), Confederate general during the Civil War. Died after being wounded at the battle of Chancellorsville.
- Franz Liszt (1811–1886), composer. Died of pneumonia caught while attending a music festival.
- Leo Tolstoy (1828–1910), novelist. Died of pneumonia at a railway station following exposure to cold.
- Harriet Tubman (c. 1820–1913), former slave and leader of the Underground Railroad. Died of pneumonia following several years of declining health.
- Bert Lahr (1895–1967), actor who played the Cowardly Lion in *The Wizard of Oz*. Died of pneumonia while making another film.
- Jim Henson (1936–1990), puppeteer best known for the Muppets movies and



Harriet Tubman. COURTESY OF THE LIBRARY OF CONGRESS.

television show. Died of multiple organ failure following bacterial pneumonia.

- Luciano Pavarotti (1935–2007), opera singer. Died of pneumonia as a complication of treatment for pancreatic cancer.
- Emerging diseases. Some emerging diseases, such as severe acute respiratory syndrome (SARS) and avian influenza (bird flu) can cause pneumonia in otherwise healthy people.

# Description

Pneumonia is one of the oldest diseases known to humans, first described by Hippocrates in the fifth century *BCE*. Pneumonia can vary from a mild infection of the lungs that does not require hospital treatment to a fatal illness. In the 1930s pneumonia was the leading cause of death in the United States. It is still the eighth most common cause, killing 60,000 people each year. Pneumonia can come on either suddenly or gradually. Its major symptoms include chest pain, fever, severe coughing, and greenish or pus-colored sputum (mucus or phlegm). The early stages of pneumonia are sometimes mistaken for a cold or flu. The severity of the symptoms depends on the organism causing the illness and the person's basic level of health.

Bacterial pneumonia often comes on suddenly with sweating, severe chest pain, high fever (up to 105°F/40.6°C), chills, and a cough that produces greenish or yellowish sputum. It can develop by itself or following a viral infection like a cold or the flu.

Viral pneumonia accounts for about half of all cases. Some cases are mild. In other cases, people have symptoms resembling those of influenza: fever, aching muscles, headache, and weakness. Within twelve to thirty-six hours, however, patients become much sicker, may start gasping for breath, and their cough becomes worse. Patients with viral pneumonia sometimes develop a secondary bacterial pneumonia.

Pneumonia caused by mycoplasma is usually mild and develops gradually. It is common among children and young adults because it spreads rapidly in day care, college dormitories, and other group settings. The most noticeable symptoms of mycoplasmal pneumonia are a dry cough and whitish sputum. Some patients also experience nausea and vomiting.

## **Demographics**

Pneumonia is a widespread health problem in the general population as well as hospital inpatients and those in nursing homes. According to the Centers for Disease Control and Prevention (CDC), 1.4 million people in the United States are treated in hospitals each year for pneumonia, with an average stay of five days. About 2 percent of nursing home residents are treated for pneumonia in an average year.

Pneumonia is most common in the winter months in the United States. It affects males more often than females, and African Americans more often than members of other racial or ethnic groups.

In developing countries, pneumonia is a common cause of death among children. The World Health Organization (WHO) estimates that 2 million children die each year around the world, and that one in three deaths among newborn babies results from pneumonia. People who are at increased risk of pneumonia include:

- Those with weakened immune systems, including people who are HIV-positive, have received organ transplants, or are being treated for cancer with chemotherapy
- People with emphysema, cystic fibrosis, or other diseases that affect the lungs
- People who smoke or abuse alcohol
- People who are hospitalized or in nursing homes, particularly those placed on artificial respirators
- People whose jobs expose them to pesticides, large amounts of dust, or other chemicals that irritate the lungs
- Very young children and adults over age sixty-five
- People with diabetes, kidney disorders, sickle cell anemia, or heart disease

# **Causes and Symptoms**

The causes, types, and major symptoms of pneumonia have been described in previous sections.

Other symptoms that some people with pneumonia experience include:

- Rapid but shallow breathing
- Loss of appetite
- Mental confusion and disorientation (more common in the elderly)
- Unusual tiredness
- Heavy sweating and clammy skin

# Diagnosis

The diagnosis of pneumonia is made by a combination of the patient's history, a physical examination, and appropriate laboratory and imaging tests. The doctor will need to know when the symptoms started, whether the patient has been recently hospitalized, whether he or she has had a cold or flu, and similar questions. The physical examination will include listening to the patient's breathing through a stethoscope as well as taking the temperature and pulse. Patients with pneumonia typically have abnormal rubbing, crackling, or other harsh sounds that the doctor can hear. In addition, the number of breaths per minute is usually high. A chest x ray will usually be ordered. Other tests include:

- A complete blood count (CBC). An unusually high number of white blood cells is a common sign of an infection.
- A sputum test. The doctor can collect a sample of the patient's sputum and send it to a laboratory to identify the organisms that may be causing the infection. Not everyone with pneumonia will produce sputum and not everyone who produces sputum has pneumonia.

### Treatment

Treatment depends on the cause of the pneumonia. Pneumonia caused by bacteria or mycoplasma is treated with a seven- to ten-day course of antibiotics. Viral infections cannot be treated by antibiotics; in some cases, the doctor may prescribe antiviral medications like rimantadine, which is used to treat some types of influenza. Most patients can care for themselves at home by taking the prescribed medication and by getting plenty of rest, drinking fluids, and taking over-the-counter pain relievers to reduce fever and headaches. It is important for people recovering at home not to return to work or a normal schedule of activities too quickly, as the pneumonia can recur. Recurrences can be more severe than the initial infection.

People who are severely ill and having difficulty breathing are hospitalized, given higher levels of oxygen to breathe, and given intravenous antibiotics. They will be given a follow-up x ray by the doctor after they are well enough to leave the hospital.

## Prognosis

The prognosis for pneumonia depends on the organism or irritant causing it, the patient's age and general health, and the time elapsed before diagnosis and treatment. Young people who are diagnosed and treated early may recover in about ten days. Middle-aged adults may take several weeks to recover fully. Mycoplasmal pneumonia takes somewhat longer than either bacterial or viral pneumonia to go away—sometimes as long as six weeks.

The prognosis for recovery is poor for people who develop ventilatorassociated pneumonia (about 50 percent will die) or those with bacterial pneumonia that spreads into the bloodstream (about 20 percent will die). Doctors sometimes use a rule called the pneumonia severity index or PSI to estimate a specific patient's chances of recovery. The PSI is based on age, gender, vital signs, and laboratory test results.

#### Prevention

An important measure that anyone can take to lower the risk of pneumonia is to quit smoking (or not start in the first place). Another step is vaccination. There is a specific vaccine called Pneumovax that protects against a specific bacterium called the pneumococcus, a common cause of bacterial pneumonia. Pneumovax is recommended for children under two and older children with sickle cell anemia, diabetes, or other diseases that affect the immune system, as well as for adults over fifty-five. A different version of this vaccine, called Prevnar, is now given to children. Older adults should also receive yearly flu shots, as bacterial pneumonia is a common complication of influenza.

### The Future

Pneumonia is likely to continue to be a common health problem, especially since it has so many different causes. On the one hand, the development of effective vaccines against flu viruses and the pneumococcus has had some positive effects in reducing the number of cases among children and older adults in the United States. On the other hand, the development of drug-resistant bacteria and the emergence of such new diseases as severe acute respiratory syndrome (SARS) present new challenges to doctors and public health officials.

**SEE ALSO** AIDS; Avian influenza; Common cold; Emphysema; Influenza; Parkinson disease; Severe acute respiratory syndrome

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# WORDS TO KNOW

- Alveoli (singular, alveolus): Tiny air sacs in the lungs where carbon dioxide in the blood is exchanged for oxygen from the air.
- **Aspiration:** The entry of food, liquids, or other foreign substances into the lungs during the breathing process.
- **Community-acquired:** Referring to a disease that a person gets in the course of ordinary activities rather than in a hospital or clinic.

- **Mycoplasma:** A very small bacterium that causes a mild but long-lasting form of pneumonia.
- **Nosocomial:** Referring to a disease that a person gets while hospitalized.
- **Opportunistic infection:** An infection that occurs only in people with weakened immune systems.
- **Sputum:** Mucus and other matter that is coughed or brought up from the lungs or throat.
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# Polio

# Definition

Polio is an infectious disease caused by a virus that normally lives in the human digestive tract. About 90 percent of persons infected by the virus have no symptoms at all; in the other 10 percent, the polio virus causes symptoms ranging from a mild flu-like illness to paralysis of the lower limbs or death from paralysis of the muscles that control breathing.
## Description

Polio was a greatly feared disease in the early part of the twentieth century because of epidemics that used to occur in the summer and fall of every year. Ironically, these epidemics were a byproduct of the improved sanitation and standards of cleanliness in large cities in Europe and the United States toward the end of the nineteenth century. Although polio had been known since ancient Egypt, the disease was relatively uncommon. The reason for this was that most young children before the nineteenth century were exposed to the polio virus early in life and thereby acquired lifetime immunity. Better hygiene, however, meant that infants and young children had fewer opportunities to encounter and develop immunity to polio. Exposure to poliovirus was therefore delayed until later in childhood or adult life, when it was more likely to cause paralysis or death.

The first recorded outbreak of polio in the United States occurred in Louisiana in 1841. Outbreaks were few until the early 1900s, however, when large cities like New York and Boston began to experience annual polio epidemics. In 1916, 27,000 people across the United States fell ill with polio and 6,000 died. The epidemics of the 1940s and 1950s were even worse; the 1952 epidemic was the worst in the country's history. There were 58,000 cases reported; 3,145 people died, and 21,000 were left paralyzed. This history helps to explain why the introduction of the Salk vaccine in 1955 was regarded as a miracle. By 1957, the success of the vaccine was reflected in the fact that the annual number of cases fell to 5,600. After the development of the Sabin oral vaccine in the early 1960s, the annual number of polio cases in the United States fell to 160. The last cases of endemic polio in the United States were reported in 1979.

There are four major categories of polio:

- Asymptomatic: Between 90 and 95 percent of people infected by the polio virus have no symptoms at all.
- Minor flu-like illness: Between 4 and 8 percent of patients. This minor illness is sometimes called abortive polio.
- Nonparalytic meningitis: This is a condition that affects about 2 percent of patients. It is characterized by headache; pain in the neck, back and abdomen; fever; vomiting; and irritability.
- Paralytic polio: Between one in 200 and one in 1,000 patients develop a paralytic polio.

#### Also Known As

Poliomyelitis, infantile paralysis

Cause

Poliovirus

#### Symptoms

May range from none at all to paralysis and death

#### Duration

A few days to several months; complications may occur years later

## Demographics

Polio was widespread in the developed countries in the first part of the twentieth century. The epidemics not only became more severe, but also affected adolescents and adults rather than mostly children. The older average age of patients was also marked by increased severity of symptoms. Since the introduction of effective vaccines, paralytic polio is almost unknown in the United States except among recent immigrants and other groups (such as the Amish) that do not routinely participate in community-wide vaccination programs.

Worldwide, polio epidemics are most common in tropical countries during the months of July through September. Both sexes and all races are equally likely to get the disease if they are not protected by immunization.

Some people are more likely than others to develop the paralytic form of the disease if they do become infected:

- Young children
- Elderly adults
- People who engage in hard physical labor or strenuous exercise
- · People who have recently had a tonsillectomy or dental surgery
- Pregnant women
- · People who travel frequently to areas where polio is still endemic
- Those with an immune system weakened by HIV or certain types of cancer treatment

## **Causes and Symptoms**

Polio is caused by a virus that enters the mouth through food or water that has been contaminated by fecal matter. It is a contagious illness; anyone living with a recently infected person can become infected. Although people carrying the poliovirus are most contagious for seven to ten days before and after symptoms (if any) appear, they can spread the virus for weeks in their bowel movements.



*Two girls with polio at a special school in New Delhi, India.* AP IMAGES.

## **Defeating Polio**

Many people think of Franklin Delano Roosevelt (1882–1945) as the individual whose struggle called public attention to the need to find a vaccine against polio. Roosevelt acquired polio in August 1921 while vacationing with his family in New Brunswick, Canada. After falling from his boat into the cold waters of the bay on August 9, Roosevelt went swimming and jogging the next day with his children. He felt unwell that night; two days later, his legs were completely paralyzed. In spite of a variety of treatments, Roosevelt remained permanently paralyzed from the waist down until his death in 1945.

Roosevelt refused to allow his condition to prevent him from serving as governor of New York and later as president of the United States. He was instrumental in founding the National Foundation for Infantile Paralysis, later the March of Dimes. Roosevelt's foundation supported the work of Dr. Jonas Salk (1914–1995), the researcher who developed the first safe and effective polio vaccine. Salk's vaccine was licensed in 1955 and was followed by mass vaccination campaigns across



President Franklin D. Roosevelt.

the United States. The Salk vaccine, reformulated for additional potency in 1987, is currently the standard polio vaccine used in the United States.

Once inside the body, the polio virus takes between six and twenty days to incubate. It finds its way to the tissues lining the throat and the intestinal tract, where it multiplies rapidly. After about a week in the intestines, the virus travels to the tonsils and the lymph nodes, where it multiplies further and then enters the bloodstream. In a minority of cases, the virus enters the central nervous system (CNS) from the blood and lymph. It then multiplies in and destroys the nerve cells in the brain known as motor neurons that control the movements of the muscles. The location and severity of the paralytic polio that results when the motor neurons are damaged varies with the part of the CNS that is affected.

Between 4 and 8 percent of polio infections are characterized by influenza-like symptoms known as abortive poliomyelitis. People with

this form experience sore throat and fever, nausea, vomiting, abdominal pain, constipation, or diarrhea. Abortive polio is difficult to distinguish from the flu or other viral infections. Patients recover completely in about a week.

Patients with nonparalytic meningitis may experience a brief period of general illness followed by stiffness in the neck, back, or legs. They may also experience other abnormal sensations for a period of two to ten days. As with abortive polio, patients with nonparalytic meningitis recover completely.

Paralytic polio is usually divided into three types, depending on whether the paralysis affects the arms and legs (spinal polio; accounts for 79 percent of cases of paralytic polio); breathing, speaking, and swallowing (bulbar polio; 2 percent of cases); or the limbs as well as breathing and other functions (bulbospinal polio; 19 percent of cases). Bulbar polio is particularly likely to lead to death if the patient is not placed on a respirator because the virus affects the brain stem—the part of the brain that controls heartbeat as well as breathing and other vital functions.

#### Diagnosis

The diagnosis of polio is based on a combination of the patient's history and the type and location of symptoms—particularly such symptoms as a stiff neck, difficulty breathing, or abnormal reflexes. To confirm the diagnosis, samples of the patient's stool, spinal fluid, or throat mucus may be collected and sent to a laboratory for analysis to see whether the sample contains the virus itself. A blood sample early in the infection may also be analyzed for evidence of antibodies to the poliovirus.

#### Treatment

There is no cure for polio. Patients with abortive polio or nonparalytic meningitis do not usually need treatment other than resting at home.

Patients with paralytic polio may be placed on a respirator to help them breathe, particularly if they are diagnosed with bulbar polio. Other treatments include painkillers and hot packs for muscle aches, physical therapy to restore muscle strength, and occupational or speech therapy as needed. Braces or special shoes may be recommended for some patients. A few patients may undergo surgery to restore limb function. The overall prognosis for recovery from an acute attack of paralytic polio is generally good. Mortality is about 5–10 percent, mostly in elderly and very young patients. Half the patients with spinal polio recover fully; 25 percent have mild disabilities; and the remaining 25 percent are left with severe disabilities. Most patients recover from breathing problems, and only a small percentage of patients need long-term treatment on a respirator. Patients with muscle paralysis typically recover about 60 percent of their strength in the first three to four months of treatment.

Some patients who have recovered from paralytic polio develop a disorder called post-polio syndrome (PPS) between ten and forty years after the initial infection. PPS is not a reinfection, although its cause is not completely understood. PPS is marked by:

- Muscular weakness
- Fatigue
- Being easily exhausted after even small amounts of activity
- Joint pain
- Sleep disorders
- Difficulty breathing or swallowing
- Inability to tolerate cold temperatures

PPS is treated with rest and such supportive measures as powered wheelchairs, pain relievers, and medications to help the patient sleep. Patients are also encouraged to simplify their work habits and take frequent rest breaks.

## Prevention

Polio can easily be prevented by administration of either the Salk vaccine, which contains an inactivated poliovirus, or the Sabin oral vaccine, which contains a weakened live virus. The Salk vaccine is usually given in two doses four to eight weeks apart followed by a third dose six to twelve months after the second dose. It is the only polio vaccine that is given to people with weakened immune systems.

The Sabin vaccine is given in a single dose by mouth. It is not routinely given to people with weakened immune systems.

## The Future

There is hope that polio will follow smallpox as a disease that humankind has completely wiped out. In 1994 both North and South America were

## WORDS TO KNOW

Asymptomatic: Having no symptoms.

**Endemic:** A term applied to a disease that maintains itself in a particular area without reinforcement from outside sources of infection.

**Meningitis:** Inflammation of the membranes that cover the brain and spinal cord.

**Motor neuron:** A type of cell in the central nervous system that controls the movement of muscles either directly or indirectly.

declared polio-free, followed by Australia, Japan, China, and other countries around the Pacific Ocean in 2000, and Europe in 2002. As of 2008, however, there were still four countries in the world where polio was endemic: Nigeria, India, Pakistan, and Afghanistan. Public health doctors hope that the vaccination campaigns that are underway in these countries will clear them of the disease in the next few years.

SEE ALSO Chronic fatigue syndrome; Influenza; Sleep apnea

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# m

## Postpartum Depression

## Definition

Postpartum depression, or PPD, is a mood disorder that occurs in women following childbirth. It is more serious than the so-called baby blues. The baby blues are a mild mood disturbance that affects most women for a few days or a week after childbirth, goes away on its own, and does not interfere with the mother's ability to care for the baby. In PPD, the depressed feelings are more intense, last longer, and usually affect the mother's care giving. In a very few cases, the mother develops a condition called postpartum psychosis shortly after childbirth. Postpartum psychosis is a severe mental disorder in which the mother suffers from delusions or hallucinations and may harm herself or the baby.

## Description

Postpartum depression usually develops gradually over a period of weeks after the baby's birth. Unlike the baby blues, PPD does not go away by itself after the first two weeks after childbirth. The new mother typically begins to have difficulty sleeping, feels tired and tearful much of the time, may worry a lot about her ability to take care of the baby, loses interest in activities that she used to enjoy, may have difficulty in nursing or otherwise caring for the baby, may feel angry with her husband, and may draw away from friends and family.

Postpartum psychosis, which is rare, has a different pattern of onset; it often develops suddenly. A woman with postpartum psychosis begins to have hallucinations and delusions (false beliefs) within about three days of the baby's birth. She may believe, for example, that the baby is

Also Known As PPD, postnatal depression

#### Cause

Hormonal and psychosocial factors, history of depression

#### Symptoms

Anxiety, irritability, headaches, intense mood swings, tearfulness, inability to function

#### Duration

Months to years

Satan or is going to die shortly, or she may hear voices telling her to hurt or kill the child. The women most at risk for postpartum psychosis are those with a history of bipolar disorder.

## Demographics

The American College of Obstetricians and Gynecologists estimates that between 70 and 80 percent of new mothers experience the baby blues after childbirth, and about 10 percent suffer postpartum depression. Postpartum psychosis occurs in one or two new mothers per thousand.

Some women are at greater risk than others of having postpartum depression:

- Women with a personal history of depression.
- Women with a previous episode of postpartum depression.
- Women diagnosed with depression during pregnancy.
- Single mothers.
- Women whose pregnancy was unwanted or unplanned.
- Women who have suffered recent losses, such as a death in the family, unemployment, or a traumatic accident.
- Women who were abused by their mothers or had difficult relationships with them.
- Women who are unhappily married or have little social support.
- Women whose income is low; 24.3 percent of women with annual incomes below \$10,000 develop PPD compared to 10.8 percent whose income is \$50,000 or higher.

## The Andrea Yates Case

Andrea Yates (1964–) made headlines across the United States in June 2001 when she drowned her five children in a bathtub in her house in Texas. Yates had married her husband in 1993 with the expectation of "having as many babies as nature allowed." In July 1999, after four children and one miscarriage, Yates collapsed, attempted suicide twice, and was taken to a psychiatric hospital. There she was diagnosed with postpartum depression and psychosis. Her first psychiatrist urged her not to have any more children due to the risk of another psychotic depression. Nevertheless, Yates and her husband conceived their fifth child one month after Andrea was discharged from the hospital in January 2000.

Yates's postpartum psychosis recurred in March 2001, three months after the birth of her last child and a few days after her father-in-law's death. At the beginning of April, she became unable to function at all and was hospitalized again. Although Yates's husband was advised by a new psychiatrist not to leave her alone, he went to work on June 20, an hour before his mother was to come to the house to help Andrea with the children. In the space of that hour, Yates drowned all five children in the bathtub. Although Yates was convicted of murder in 2002, in 2006 she received a new trial and was found not guilty by reason of insanity. As of 2008 she is in a low-security state mental hospital in Texas. Although the Yates case reopened a long-standing disagreement about using insanity as a legal defense, it also made many more people aware of the existence and possible consequences of postpartum depression and psychosis.

- Women with low self-esteem.
- Women whose infant is colicky, temperamental, has birth defects, or is otherwise difficult to care for.

One study of 27,000 women done in 2006 reported that the rate of postpartum depression varies by race or ethnicity even when factors like income, education level, age, marital status, and the child's health are accounted for. The researchers found that 15.7 percent of their sample suffered from PPD. Of the depressed group, 25.2 percent were African American, 22.9 percent were American Indian/Native Alaskan, 15.5 percent were Caucasian, 15.3 percent were Hispanic, and 11.5 percent were Asian/Pacific Islander.

## **Causes and Symptoms**

Doctor have proposed several different explanations for PPD.

- Changes in hormone levels. After childbirth, the levels of estrogen, progesterone, and thyroid hormone in the woman's body drop sharply within forty-eight hours. Some women appear to be more sensitive to these changes than others, and may therefore develop PPD.
- Other physical changes. Many women feel tired, physically unattractive, and worried about their weight after pregnancy. Many also feel pain in the area around the vagina, or in the abdomen if the baby was delivered by cesarean section.
- Mood changes caused by interrupted sleep and responsibilities related to caring for the baby. Some new mothers feel trapped at home or feel a loss of identity, particularly if they had been working during pregnancy.
- Believing certain myths about "being a perfect mother" or "having a perfect baby." Women who have unrealistic expectations of motherhood—for example, that they should never lose their temper with the baby—are more likely to feel depressed when they cannot live up to impossible standards.

In addition to tearfulness, anxiety, and fatigue, women with PPD may have the following symptoms:

- Headaches
- Loss of interest in sex
- Extreme mood swings

- Either excessive concern about the baby or at the other extreme, lack of interest in the baby
- Weight loss
- Feelings of guilt or worthlessness
- Feelings of rejection
- Thoughts of suicide or death
- Insomnia and other sleep disturbances
- Difficulty thinking or concentrating
- Inability to enjoy activities that used to give pleasure, satisfaction, or feelings of accomplishment

## Diagnosis

After taking a personal and family history of depression and other mood disorders, the doctor will usually give the patient a blood test to make sure that the moodiness and other symptoms are not caused by an underactive thyroid gland. In addition, the doctor will ask the patient to fill out a questionnaire called the Edinburgh Postnatal Depression Scale. It is a short list of ten questions. A score of twelve or higher, or a "yes" answer to the last question (about suicidal thoughts), indicates that the patient needs further treatment for her depression.

## Treatment

Treatment for postpartum depression usually consists of a combination of antidepressant medications and psychotherapy. The medications must be carefully chosen if the mother is breastfeeding because they will be passed on to the baby in the mother's milk. Some antidepressants will not cause problems for the baby but others have not yet been tested for safe use by nursing mothers.

Cognitive behavioral therapy is often recommended for new mothers who feel depressed or incompetent because they have had unrealistic ideas of what it takes to be a "good" mother or what babies are really like. Many women find support groups for new mothers helpful too.

Some women with PPD benefit from short-term hormone therapy. Estrogen replacement can sometimes improve mood; its drawbacks, however, include decreased production of breast milk and an increased risk of blood clots in the legs. Other women may need treatment with thyroid hormone. Women with postpartum psychosis require treatment in a hospital as this condition is considered a psychiatric emergency. They are usually given a combination of mood stabilizers, tranquilizers, and antipsychotic drugs.

## Prognosis

Most women with PPD start to feel better within two to four weeks of starting treatment, although complete recovery may take months. If the woman has not had a previous episode of depression, six to twelve months of treatment is recommended.

## Prevention

Some researchers think that a high-protein diet during pregnancy and taking supplements containing vitamins and omega-3 fatty acids help to lower the risk of postpartum depression. Omega-3 fatty acids are found naturally in such fish as tuna and salmon, and in some nuts. They play a role in proper brain functioning.

Early treatment of PPD is critical as it lowers the risk of a recurrence with later pregnancies as well as improving the mother's relationship with her child. A woman who is not treated for PPD has a 90 percent chance of having the illness recur with her next pregnancy.

## The Future

Adequate recognition and treatment of PPD are important to the wellbeing of the children of these distressed mothers. Since postpartum depression interferes with a mother's ability to care for her baby, it can lead to serious long-term later problems in the mother/child relationship. About 4 percent of mothers with postpartum psychosis who are not treated end up killing their children.

**SEE ALSO** Bipolar disorder; Child abuse; Depression

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## WORDS TO KNOW

- **Baby blues:** An informal term for the temporary sad feelings some mothers feel for a week or so after childbirth. It is less serious than postpartum depression and usually goes away by itself.
- **Delusion:** A false belief that a person maintains in spite of evidence to the contrary, such as that one's baby is a child of God or the devil.

Estrogen: A female hormone produced in the ovaries.

- Hallucination: Seeing things or hearing voices in the absence of genuine stimuli.
- **Postpartum:** Referring to the period of time after giving birth.
- **Postpartum psychosis:** A severe mental disorder in which the mother suffers from delusions or hallucinations.

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## m

## Posttraumatic Stress Disorder

## Definition

PTSD is an anxiety disorder that develops after a frightening ordeal involving physical harm or the threat of physical harm or death. First diagnosed in soldiers, it is now recognized in civilian survivors of rape or

#### Posttraumatic Stress Disorder

Firefighters outside a fire station that was heavily damaged in the September 11, 2001 terrorist attacks. Many first responders suffered from PTSD after their involvement in the rescue and recovery efforts. AP IMAGES.



other criminal assaults, natural disasters, plane crashes, train collisions, industrial explosions, acts of terrorism, child abuse, or war.

## Description

The experience of PTSD has sometimes been described as like being in a horror film that keeps replaying and cannot be shut off. It is common for people with PTSD to feel intense fear and helplessness, and to relive the frightening event in nightmares or in their waking hours. Sometimes the memory is triggered by a sound, smell, or image that reminds the sufferer of the traumatic event. This reliving of the event is called a flashback. People with PTSD are also likely to be jumpy, easily startled, or to go numb emotionally and lose interest in activities they used to enjoy. They may have problems with memory and with getting enough sleep. In some cases they may feel disconnected from the real world or have moments in which their own bodies seem unreal. Many people with PTSD turn to alcohol or drugs in order to escape the flashbacks and other symptoms, even if only for a few minutes.

## Demographics

PTSD can develop in almost anyone in any age group exposed to a sufficiently terrifying event or chain of events. The National Institute of Mental Health (NIMH) estimated in 2007 that about 7.7 million adults in the

Also Known As PTSD

#### Cause

Exposure to a frightening or horrifying event or series of events

#### Symptoms

Flashbacks, emotional numbing, jumpiness, avoidance of people or places linked to the event

#### Duration

Months to years

## A Quiet Hero's Struggles with PTSD

Roméo Dallaire (1946–), a member of the Canadian Senate, formerly commanded the United Nations peacekeeping mission in Rwanda in 1994. Although Dallaire made repeated requests to receive more troops to prevent mass murder during the violent civil war, he and his troops could not stop thousands of Rwandans from being massacred. Dallaire later described the horrors he witnessed in *Shake Hands with the Devil* (2003).

After returning to Canada, Dallaire developed signs of depression but received other commands in the Canadian Army. He began having flashbacks, which he later described in an interview he gave in 2002: "Your mind with time, in fact, doesn't erase things that are traumas. It makes them clearer. They become digitally clearer and then you are able to sit back and all of a sudden have every individual scene come to you."

Dallaire was diagnosed with PTSD in 2000 and given a medical retirement from the Canadian Army. His PTSD continued to get worse, causing him to drink too much and have thoughts of suicide. In June 2000 he was found in an alcoholic coma. After leaving the hospital, he decided that writing and lecturing about the genocide he had witnessed was the key to recovery. Dallaire became active in many humanitarian causes, including helping children affected by war and becoming a member of a research center for the study of genocide. He credits his family and his religious faith with giving him the support he needed when his traumatic memories almost overwhelmed him.



Canadian General Roméo Dallaire. AP IMAGES.

United States have PTSD. One study found that 3.7 percent of a sample of teenage boys and 6.3 percent of adolescent girls had PTSD. It is estimated that a person's risk of developing PTSD over the course of his or her life is between 8 and 10 percent. On average, 30 percent of soldiers who have been in a war zone develop PTSD. Women are at greater risk of PTSD following sexual assault or domestic violence, while men are at greater risk of developing PTSD following military combat.

PTSD is more likely to develop after an intentional human act of violence or cruelty such as a rape or mugging than as a reaction to an impersonal catastrophe like a flood or hurricane.

PTSD can develop in therapists, rescue workers, or witnesses of a frightening event as well as in those who were directly involved.

## **Causes and Symptoms**

The causes of PTSD are not completely understood. It is unknown why some people involved in a disaster develop PTSD and other survivors of the same event do not. For example, after terrorists destroyed the World Trade Center in New York City on September 11, 2001, a survey was conducted in November of 988 adults living close to the site. Researchers found that only 7 percent had been diagnosed with PTSD following the events of September 11; the other 93 percent were anxious and upset, but did not have PTSD. One theory proposed by a neurobiologist is that people who develop PTSD have lower blood levels of cortisol, a stress hormone, even before the traumatic event. Cortisol appears to prevent or minimize the imprinting of frightening memories involved in flashbacks. Another theory is that trauma causes changes in certain parts of the brain involved in the processing of memory and emotion.

Factors that influence the severity of PTSD include:

- The nature, intensity, and duration of the traumatic experience. For example, someone who just barely escaped from the World Trade Center before the towers collapsed is at greater risk of PTSD than someone who saw the collapse from a distance or on television.
- The person's previous history. People who were abused as children, who were separated from their parents at an early age, or who have a previous history of anxiety or depression are at increased risk of PTSD.
- Genetic factors. Vulnerability to PTSD is known to run in families.

• The availability of social support after the event. People who have no family or friends are more likely to develop PTSD than those who do.

The symptoms of PTSD usually emerge within three months of the frightening event, although in some cases they may take several years to develop. A person must have the following symptoms for at least a month to be diagnosed with PTSD:

- Flashbacks
- Difficulty sleeping; having nightmares about the event
- Trying to avoid reminders of the event
- Emotional numbness, inability to enjoy previously pleasurable activities
- Anger and irritability
- Memory problems and having difficulty concentrating
- Being unusually jumpy and easily startled; this type of symptom is called hyperarousal
- Intense feelings of shame or guilt
- Feelings of unreality, such as someone feeling that one's body isn't real or that the outside world isn't real
- Hopelessness about the future
- Self-destructive behavior, such as drinking too much or taking drugs

## Diagnosis

The diagnosis of PTSD is based on the patient's history, including the timing of the traumatic event and the duration of the patient's symptoms. There are no laboratory or imaging tests that can detect PTSD.

## Treatment

Treatment for PTSD usually involves a combination of medications and psychotherapy. If patients have started to abuse alcohol or drugs, they must be treated for the substance abuse before being treated for PTSD. The medications are given to help patients sleep better, to improve their memory and ability to concentrate, and to feel less irritable or fearful. In addition to tranquilizers and antidepressants, some drugs that were originally developed to treat epilepsy appear to help some patients with PTSD. The doctor may need to try several different types of medications before finding the one that works best for an individual patient. The types of psychotherapy that may be used include individual therapy, group therapy, family therapy, and relaxation techniques. Some patients are also helped by hypnosis, art therapy, pet therapy, or music therapy. Patients with PTSD are not hospitalized unless they are threatening to commit suicide or harm other people.

Alternative therapies that have been reported to help people with PTSD are acupuncture, therapeutic massage, meditation, and prayer. In addition, Native Americans are often helped to recover by participating in traditional tribal rituals for cleansing memories of war and other traumatic events.

#### Prognosis

The prognosis of PTSD is difficult to determine because patients' personalities and the experiences they undergo vary widely. A majority of patients get better, including some who do not receive treatment. One study reported that the average length of PTSD symptoms in patients who get treatment is thirty-two months, compared to sixty-four months in patients who are not treated.

Factors that improve a patient's chances for full recovery include prompt treatment, early and ongoing support from family and friends, a high level of functioning before the frightening event, and an absence of alcohol or substance abuse.

About 30 percent of people with PTSD never recover completely, however. A few commit suicide because their symptoms get worse rather than better.

#### Prevention

PTSD is impossible to prevent completely because natural disasters and human acts of violence will continue to occur. In addition, it is not possible to tell beforehand how any given individual will react to a specific type of trauma. Prompt treatment after a traumatic event may lower the survivor's risk of developing severe symptoms.

#### The Future

It is possible that further research into the effects of trauma on memory and other mental functions will lead to new options in treating PTSD.

**SEE ALSO** Gulf War syndrome; Stress

## WORDS TO KNOW

**Cortisol:** A hormone produced by the adrenal glands near the kidneys in response to stress.

- **Flashback:** A temporary reliving of a traumatic event.
- **Hyperarousal:** A state of increased emotional tension and anxiety, often including jitteriness and being easily startled.
- **Trauma:** A severe injury or shock to a person's body or mind.

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veterans. It includes twelve short videos, including Jim Northrup's recitation of his poem "Walking Point" at http://www.wpt.org/wayofthewarrior/video. cfm?id=150. Northrup, a member of the Fond du Lac Anishinaabe tribe and a Vietnam veteran, turned to writing as a way of coping with PTSD.

## **m** Prematurity

## Definition

Prematurity or preterm birth refers to the birth of a baby before the thirty-seventh week of pregnancy. A normal pregnancy lasts between thirty-eight and forty-two weeks after conception. Babies born after forty-two weeks are called post-term babies.

Prematurity is a condition rather than a disease or a disorder by itself. However, babies born prematurely are at high risk of heart problems, breathing difficulties, jaundice, infections, and other health problems because their internal organs and immune systems are not yet fully developed at the time of birth.

## Description

Prematurity refers to birth before the thirty-seventh week of the mother's pregnancy. A premature or preterm baby is smaller than a full-term baby and weighs less. Premature babies (sometimes called "preemies") need special care in the hospital after they are born and frequent medical checkups in the first two years after birth because of their increased risk of health problems. A baby may be born prematurely because of the mother's lifestyle or because of factors beyond the mother's control, including her age and basic bone structure.

Doctors describe babies born prematurely in terms of their birth weight as well as their gestational age (the number of weeks the mother was pregnant before the baby was born). Although birth weight usually increases along with gestational age, some babies are larger or smaller than normal for their gestational age. Preterm babies are classified according to their weight as follows:

• Low birth weight (LBW) babies are those that weigh less than 5 pounds 3 ounces (2,500 grams) at birth.

#### Also Known As

Premature birth, preterm birth

#### Cause

Diseases or malnutrition in the mother; young age; multiple births

#### Symptoms

(In mother) Contractions, backache, spotting, other signs of early delivery

#### Duration

Complications may last well into childhood

#### Prematurity



**Premature infant shortly after birth.** SHUTTERSTOCK.

- Very low birth weight (VLBW) babies are those that weigh less than 3 pounds 5 ounces (1,500 grams) at birth.
- Extremely low birth weight (ELBW) babies are those that weigh less than 2 pounds 3 ounces (1,000 grams) at birth. About 10 percent of babies born with low birth weight are ELBW infants.

Because a higher gestational age makes a significant difference in a baby's chances for normal health and development as well as simple survival, the doctor will try to reduce the mother's risk of giving birth prematurely whenever possible. In some cases the mother may be advised to leave a job that requires long hours of standing on her feet, to eat a healthier diet, or to quit smoking. Another method that appears to be effective in reducing the risk of preterm delivery is injecting a hormone called progesterone or applying a cream containing the hormone inside the vagina.

## **Demographics**

About 12 percent of babies born in the United States—one out of every eight—are born prematurely. Prematurity is one of the leading causes of death among newborn babies; in the United States, it accounts for about 25 percent of deaths among newborns. There are about 500,000 babies born prematurely in the United States each year; 32,000 of these are ELBW infants. Risk factors for low birth weight infants include:

- Low socioeconomic status
- African American race
- Mother's age below fourteen or over forty
- A history of giving birth to preterm infants

#### **Causes and Symptoms**

The cause of premature birth is not known in about 50 percent of cases. In the remaining half, the causes include:

- Mother's lifestyle. Drinking, smoking, using drugs, and not eating enough are factors in premature birth. Smoking alone is a factor in 15 percent of preterm births.
- Mother's age. Mothers younger than eighteen or older than thirtyfive are more likely to have premature babies.
- Multiple pregnancy. Mothers who are carrying twins or triplets are more likely to deliver prematurely.
- Abnormalities of the mother's uterus.
- Diabetes or high blood pressure in the mother.
- Extremely overweight or underweight mother.
- Mother has had great difficulty getting pregnant.
- A history of miscarriages or abortions.
- Chronic infections of the vagina.

The signs that a mother is starting to give birth prematurely include contractions of the uterus that come more often than six per hour; a discharge of fluid from the vagina; diarrhea; spotting of blood from the vagina; a backache; or a feeling of pressure or pain below the abdomen.

Babies born prematurely are at risk for a number of severe health complications in later childhood as well as during infancy. These include:

- Breathing problems due to underdeveloped lungs
- Respiratory distress syndrome
- Jaundice
- Heart problems
- Eye disorders
- Anemia
- Infections

- Bleeding into the brain
- Developmental and learning disabilities
- Cerebral palsy

## Diagnosis

Doctors can estimate a woman's risk of giving birth prematurely by using a scoring system based on her age, medical history, daily habits, socioeconomic status, and the condition of her pregnancy. About 30 percent of women with high risk scores have their babies prematurely compared to 2 percent of those with low risk scores. One complication with estimating a baby's gestational age, however, is that many women are uncertain of the date of their last menstrual period.

To diagnose whether an expectant mother is about to give birth, the doctor will use ultrasound to measure the length and width of her cervix. This part of the uterus begins to dilate, or open up, as childbirth begins. It also becomes shorter. If the mother is having contractions, the doctor may time them as well as check to see whether the membranes surrounding the baby in the uterus have started to break.

After the baby is born, the doctor will check for health problems before the baby is placed in the newborn intensive care unit:

- Blood test. This is done to detect anemia and to evaluate the possibility that the baby will develop jaundice.
- Chest x ray. This is done to check the condition of the newborn's lungs.
- Spinal tap. This test may be done if the baby shows signs of an infection.

## Treatment

Treatment for prematurity may include trying to stop the birth process. There are several approaches that may be used. Depending on the mother's stage of pregnancy and how far the birth has progressed, bed rest may be enough to stop the contractions. Other approaches include giving the mother medications that stop the contractions or by putting stitches in the cervix (the neck of the uterus) to hold it closed until the last month of pregnancy, when the stitches are removed. Recent research indicates that giving progesterone, a female hormone, to pregnant women who have had a previous preterm baby reduces their risk of having their new baby born prematurely.

The specific treatments given to the baby depend on its weight at birth and its gestational age. As the care of premature newborns has improved since 1960, the lower limit of viability (the age at which 50 percent of premature infants will survive) has dropped from twentyeight weeks to twenty-three to twenty-four weeks. The earliest premature baby known to survive is a Canadian boy who was born twenty-one weeks and five days after conception in 1987.

All premature babies are taken to a newborn intensive care unit or NICU. Care given usually includes:

- Being placed in an incubator or warmer. Preterm babies cannot regulate their own body temperature as well as full-term babies because they do not have as much body fat. VLBW babies have very thin skin in addition to little body fat.
- Surfactant. Surfactant is a substance secreted by cells in the lungs that keeps the lungs inflated when a person breathes in and prevents the lungs from collapsing when the person breathes out. Babies born before thirty-two weeks do not have enough surfactant in their lungs to breathe easily, so the doctor will administer some surfactant.
- Monitoring of breathing. Preterm babies often suffer from apnea, or temporary stopping of breathing, because the part of the brain that normally controls breathing is not fully mature. Infants who develop apnea can be put on a ventilator or in some cases given a medication to keep the airway open.
- Administration of fluids. Preterm babies lose more water through their skin than full-term babies, so fluids are administered intravenously to keep them from dehydrating.
- Special formula for nutrition. Preterm babies need to be fed intravenously for a few days after birth because their ability to suck directly from the breast or bottle is not fully developed. After the first few days, breast milk can be pumped from the mother and fed to the baby through a tube placed in the mouth that goes down into the stomach. Since a premature baby needs more calories than a full-term baby would in order to grow, extra fortifiers or vitamins and minerals may be added to the mother's milk.

The baby is considered well enough to go home from the NICU when it can breathe without a ventilator, is steadily gaining weight, can feed directly from the mother's breast or a bottle, and does not need to

## WORDS TO KNOW

Apnea: Temporary stopping of breathing.

- **Cervix:** The neck or lowermost part of a woman's uterus that opens into the vagina.
- **Gestational age:** An infant's age at birth counting from the date of the mother's last menstrual period.
- Jaundice: A yellowish discoloration of the skin and whites of the eyes caused by increased levels of bile pigments from the liver in the patient's blood.
- **Surfactant:** A protein-containing substance secreted by cells in the lungs that helps to keep them properly inflated during breathing.

be kept warm artificially. Children born prematurely, however, will need more frequent follow-up visits to the doctor than full-term babies. They are more vulnerable to infections, difficulties learning to speak and walk, and problems with vision and hearing. The doctor may recommend that the parents take the baby to a high-risk newborn clinic or early intervention program during its first two years of life.

## Prognosis

The prognosis of premature infants depends on their age and weight at birth; the older the gestational age and the higher the birth weight, the better the baby's chances of survival. The three leading causes of death in premature infants are respiratory failure, infections, and birth defects. Advances in the care of premature infants in recent years have improved their chances of survival; more than 90 percent of babies who weigh 2 pounds (900 grams) or more at birth survive. ELBW infants have a 40–50 percent chance of survival but have a high risk of complications. Girls are more likely than boys to survive very early birth (twenty-two or twenty-three weeks).

## Prevention

An expectant mother can reduce her risk of giving birth prematurely by getting regular prenatal care; eating a healthy diet; following the doctor's recommendations for chronic health conditions like diabetes and high blood pressure; avoiding tobacco, alcohol, and drugs; cutting back on work that requires standing on the feet; getting enough sleep; and managing stress. Although weekly progesterone therapy is a relatively new preventive approach, women with several risk factors for preterm delivery or a history of giving birth prematurely may want to ask their doctor about it.

## The Future

An important question that researchers are investigating is the reason for the rise in the number of premature births in the United States since 1981. In that year 9 percent of births were premature, compared to 12 percent in 2008—an increase of 31 percent. As there are no completely effective ways to prevent prematurity, an answer to this question might lead to better preventive approaches.

**SEE ALSO** Cerebral palsy; Congenital heart disease; Developmental disability; Diabetes; Hypertension

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## Progeria

See Hutchinson-Gilford syndrome.

## **u** Prostate Cancer

## Definition

Prostate cancer is a cancer of the prostate gland, an inch-long (2.5 cmlong) walnut-shaped organ in the male reproductive system that lies under the urinary bladder and surrounds the urethra, the tube that conveys urine outside the body. It is the second most common type of cancer in adult males in the United States; only skin cancer is more common.

## Description

The prostate gland is a small organ that lies in front of the rectum and below the bladder in a human male. It secretes a thin milky-looking fluid that carries sperm. The prostate surrounds the urethra, the tube that carries urine and sperm out of the penis. If the prostate becomes enlarged, it can put pressure on the urethra and cause problems during urination.

Most cases of a swollen prostate in men are *not* due to cancer but involve a condition called benign prostatic hypertrophy (BPH), a noncancerous condition in which various tissue layers in the prostate gland swell and partially close off the urethra. The symptoms of both may include frequent urination, difficulty urinating, or frequent stopping and starting during urination. The diagnostic test used to tell the difference is described below. Having BPH does not increase a man's risk of developing prostate cancer.

In general, prostate cancer develops slowly; sometimes the cancer is found only at autopsy. About 70 percent of men who die after age ninety are found to have cancerous cells in the prostate at autopsy. Prostate cancer was not recognized as a type of cancer until 1853 and was considered very rare, most likely because life expectancies were lower in the nineteenth century and there were no effective ways to detect it early until the 1980s.

Some prostate cancers, however, are very aggressive and metastasize (spread) to other parts of the body fairly rapidly. A common location for metastases from prostate cancer is the lymph nodes in the pelvic area. Tumors in this location may cause swelling in the legs or aching **Cause** Unknown

#### Symptoms

None in early stages; difficulty urinating, bloody urine, pelvic pain, bone pain (later stages)

Duration Years



Magnified image of a prostate cancer cell. © PHOTOTAKE INC. / ALAMY.

sensations in the pelvis. Beyond the pelvis, cancer may spread from the prostate to the bones, causing the bones to fracture easily as well as ache.

Prostate cancer is second only to lung cancer as a cause of cancer-related deaths in men in the United States. Since 2006, there have been an average of 27,500 deaths each year from prostate cancer in American men.

## **Demographics**

Prostate cancer affects only males and is rarely diagnosed in men younger than forty. There are on average 235,000 new cases of prostate cancer

diagnosed in the United States each year. About one man in every six will develop prostate cancer in his lifetime. According to the National Cancer Institute (NCI), as of 2008 there were about 2 million men in the United States who have had prostate cancer and were still living.

Risk factors for prostate cancer include:

- Age. The risk increases rapidly in men over fifty years of age; 50 percent of all prostate cancers occur in men over seventy-five.
- Race. African American men are twice as likely to die from prostate cancer as Caucasian men; they also develop prostate cancer earlier than men of other races. Native American men and Asian American men have the lowest risk of prostate cancer. The reason for these differences is not known.
- Family history. Men who have a father or brother who has had prostate cancer have twice the risk of developing prostate cancer than a man without a family history of the disease. According to the National Human Genome Research Institute (NHGRI), men with any of the following family histories should be particularly concerned: three or more close relatives (father, brother, or son) with prostate cancer; a relative with prostate cancer in three successive generations on either the father's or mother's side of the family; or two or more male relatives diagnosed with prostate cancer before age fifty-five.
- Geographic location. Men living in New England or the Pacific Northwest are at increased risk of prostate cancer. The reason for this difference is not yet known.

- Diet. A high-fat diet is associated with an increased risk of prostate cancer. One theory is that fat increases the production of testosterone, a male sex hormone that may stimulate the growth of cancer cells in the prostate. Vegetarian men have a very low risk of prostate cancer.
- High testosterone levels.

## **Causes and Symptoms**

The cause or causes of prostate cancer are not completely understood. It is known to run in families, and certain genes are known to increase a man's risk of developing prostate cancer. However, no genes have yet been identified as a cause. Other proposed theories include: sexually transmitted viruses, a high level of sexual activity, dietary factors, exposure to certain chemicals in the environment, and obesity. So far none of these theories have been proven.

Many men with early-stage prostate cancer have no symptoms. Those who do develop symptoms may experience one or more of the following:

- Urinary problems: difficulty urinating; flow of urine starting and stopping; needing to urinate frequently, particularly at night; pain or burning when urinating; weak flow of urine.
- Impotence (difficulty having an erection).
- Blood in the urine or semen.
- Frequent pain in the hips, thighs, or lower back.
- Pain in the abdomen.
- Unintentional weight loss.

## Diagnosis

The diagnosis of early-stage prostate cancer is sometimes made accidentally in the course of surgery for BPH or a bladder problem. In other cases, the diagnosis is made at the end of a process of screening and further testing.

Screening usually involves a digital rectal examination (DRE) and a blood test for a protein produced by the prostate, called the prostatespecific antigen test (PSA). In a DRE, the doctor inserts a gloved finger into the rectum to feel whether the prostate is enlarged. The PSA test involves drawing a small sample of blood from a vein. BPH can also cause above-normal levels of PSA, however, so a biopsy is necessary to confirm the diagnosis of prostate cancer. To perform the biopsy, the doctor inserts a small ultrasound probe into the patient's rectum to guide the insertion of a needle that removes several small samples of tissue from the prostate gland. The samples are then sent to a laboratory for examination under a microscope.

Other tests that may be used include imaging studies of the bladder, lungs, bones, or abdomen. These can be useful in staging the cancer if the tissue biopsy shows that the patient does indeed have cancer.

## Treatment

The first step in treating any kind of cancer is called staging. Staging is a description of the location of the cancer, its size, how far it has penetrated into healthy tissue, and whether it has spread to other parts of the body. Prostate cancer is classified into four stages:

- Stage I: The cancer cannot be felt when the doctor performs a digital examination of the patient's rectum and is usually found accidentally. The cancer has not spread beyond the prostate.
- Stage II. The cancer is large enough to be detected by a PSA test or digital examination, but has not spread beyond the prostate.
- Stage III. The cancer has spread outside the prostate to the seminal vesicles but has not reached the lymph nodes.
- Stage IV. The cancer has spread to the lymph nodes in the pelvis, the bones, or the lungs.

There is no one-size-fits-all approach to treating prostate cancer. Depending on the patient's age, his PSA level, his general health and life expectancy, and the stage of the cancer, one or a combination of the following treatments may be used. It is important for a patient to discuss the various treatment options thoroughly with the doctor, because all of the therapies except watchful waiting have side effects, which may include problems with urination, bleeding from the rectum, and sexual problems.

- Watchful waiting. The patient is monitored for changes in PSA levels and may be given additional DREs and tissue biopsies.
  Watchful waiting may be appropriate for elderly men in poor health or for younger men with slow-growing tumors.
- Radiation therapy. Prostate cancers may be treated by radiation from a focused external beam or by implanting tiny pellets or seeds

of radioactive material directly into the tissue of the prostate. Implantation is done under general anesthesia.

- Hormone therapy. Hormone therapy is given to block production of the male sex hormone testosterone, which stimulates the growth of prostate cancer. One type of hormone therapy blocks the body's signals to the testicles to produce testosterone, while another approach uses a different hormone to block the body's ability to use testosterone. Hormone therapy is most effective with early-stage disease.
- Chemotherapy. This approach is often used when other treatment methods have failed because of its severe side effects.
- Surgery. There are two major types of surgery that may be performed to treat prostate cancer. One is radical prostatectomy, which involves the removal of the entire prostate gland, the seminal vesicles, and nearby lymph nodes. The other procedure is called a transurethral resection of the prostate, or TURP. The surgeon inserts an instrument through the urethra and removes a portion of the prostate by means of a wire loop at the end of the instrument. TURP is used to treat early-stage prostate cancer when the tumor is blocking urine flow.
- Cryotherapy. Also known as cryosurgery, this technique involves the use of extreme cold to destroy the cancer cells. The doctor inserts a metal rod into the prostate with the guidance of ultrasound imaging. Argon gas is used to cool the rod to -320°F (-195°C); this subfreezing temperature kills the cancer cells.

## Prognosis

The prognosis of prostate cancer depends on the stage of the disease and the patient's overall health. If caught early, this type of cancer is highly treatable. According to the American Cancer Society, 92 percent of men with prostate cancer live at least five years after diagnosis, and 67 percent survive at least ten years.

## Prevention

There is no known way to prevent prostate cancer. Researchers are, however, looking into dietary measures as a way to lower risk. A low-fat diet that is high in fruits and vegetables is thought to be beneficial, as is getting regular exercise.

## WORDS TO KNOW

**Benign prostatic hypertrophy (BPH):** A noncancerous condition in which the swelling of the prostate gland squeezes the urethra and causes difficulty in urination.

**Cryotherapy:** Also called cryosurgery. The use of extreme cold to destroy cancerous tumors or other diseased tissue.

**Metastasis (plural, metastases):** A secondary tumor caused by the spread of cancer from its primary location to another part of the body.

**Prostate:** A walnut-sized gland in males that secretes seminal fluid.

Some scientists are studying whether finasteride, a drug given to control hair loss and BPH, might prevent or delay the development of prostate cancer in men over fifty-five. However, some researchers think that finasteride may produce more aggressive tumors in men who develop prostate cancer while taking the drug. More research is presently under way.

## The Future

It is likely that new drugs and new surgical techniques will continue to be developed to treat prostate cancer. One recent innovation is the use of robot-assisted surgery to perform radical prostatectomy. Researchers are also working on gene therapy as a possible approach to treatment. In addition, genetic testing to identify an individual man's risk of developing prostate cancer is expected to be available within the next few years.

**SEE ALSO** Lung cancer; Skin cancer

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# R



Genetic

Infection

Injury Multiple

Other

Unknown

**Also Known As** Hydrophobia

**Cause** Virus

#### Symptoms

Paralysis, hallucinations, agitation, inability to drink, delirium

## Duration

Two to ten days after the first symptoms



## Rabies

## Definition

Rabies is defined by the Centers for Disease Control and Prevention (CDC) as a preventable viral disease most often transmitted to humans through the bite of an infected animal.

## Description

Rabies is a zoonosis, which means that it belongs to a group of diseases that can be transmitted to humans from infected animals. For most of human history, rabies was associated with infected dogs and was uniformly fatal until Louis Pasteur's discovery of an effective vaccine in 1885. As of the early 2000s, however, dogs kept as pets are no longer the primary source of rabies in the United States and Canada, being responsible for only 5 percent of cases. Other animal species that cause greater concern to public health doctors include:

- Wolves, coyotes, jackals, foxes, and other animals in the canine family.
- Bats. Most cases of rabies in the United States in recent years were traced to infected bats; at least thirty of the thirty-nine species of North American bats have been shown to carry rabies. Bat bites are dangerous partly because they are small and may go unnoticed.

#### Rabies

Image of an animal cell with rabies virus particles (red core within blue and yellow capsule). EYE OF SCIENCE / PHOTO RESEARCHERS, INC.



Several cases have been reported of children bitten during sleep by bats that had gotten into their bedrooms.

• Raccoons and skunks. All the states along the Eastern seaboard of the United States as well as Vermont, Pennsylvania, West Virginia, and Ohio have reported rabies in these animals.

Domestic cats and ferrets are considered low-risk species because they as well as dogs can be vaccinated against rabies. Rats, mice, and other rodents are also considered even lower-risk species for carrying rabies.

## **Demographics**

Rabies causes about 30,000 to 50,000 deaths each year worldwide, according to the World Health Organization (WHO), and is probably underreported, particularly in developing countries. In the United States, about fifty people died from rabies each year until about 1940, when widespread vaccination of dogs was put into practice. Between zero and three cases of rabies are documented each year in the United States as of the early 2000s, according to the Centers for Disease Control and Prevention (CDC). Between 16,000 and 39,000 people receive post-exposure treatment for rabies each year in the United States and Canada.

## Pasteur Finds a Treatment

Rabies was a universally fatal disease until 1885, when Louis Pasteur (1822–1895), the French chemist and microbiologist, successful treated Joseph Meister (1876–1940), a nine-year-old boy who had been mauled by a rabid dog on July 4, 1885. The boy's mother brought her son to Paris on July 6. At that point Pasteur and his assistant, Emile Roux, had been experimenting with a rabies vaccine made from the dried spinal cords of infected rabbits in order to immunize domestic dogs against the disease. He had tested the vaccine on only eleven dogs.

Pasteur, not a physician, took a personal risk in giving the child the new, intried vaccine. He consulted with several physicians who examined the boy's severe wounds and urged Pasteur to go ahead with the vaccination because Meister faced certain death otherwise. Pasteur gave Meister thirteen injections of the vaccine over a ten-day period. Three months later, when it was clear that the boy was completely healthy, Pasteur reported the case to the French Academy of Medicine.



*Louis Pasteur in his lab.* COURTESY OF THE LIBRARY OF CONGRESS.

Although deaths from rabies are now rare in North America, the CDC estimates the annual cost of rabies prevention and control exceeds \$300 million.

People of all ages, both sexes, and all races are equally likely to get rabies if bitten by an infected animal.

## **Causes and Symptoms**

The disease agent that causes rabies is a virus that seeks out the tissue of the nervous system. It is, however, easily killed by drying, detergents, and ultraviolet light. The virus is most commonly transmitted in the saliva of an infected animal and enters the human body through a bite; however, people have also been infected by nasal secretions from an animal entering the body through an open cut or wound. In a few rare cases, people have contracted the rabies virus through organ transplantation.
The course of the disease can be divided into four phases:

- Incubation. The length of the incubation period usually varies from twenty to ninety days, depending on the closeness of the bite to the central nervous system; bites on the head or neck have the shortest incubation periods. In a few cases the incubation period has lasted as long as nineteen years. The body does not develop antibodies to the virus during the incubation period.
- Prodrome. Warning symptoms of infection appear. The virus moves into the central nervous system and multiples rapidly; this phase lasts from two to ten days. About half of patients experience pain or tingling at the site of the bite; most will also have headaches, fever, loss of appetite, chills, nausea, diarrhea, insomnia, anxiety, and depression.
- Acute phase. This phase lasts from two to seven days. The patient may hallucinate, try to bite others, thrash around, and have seizures. Spasms of the throat muscles prevent swallowing water even though the patient is thirsty; he or she may drool large amounts of saliva, sometimes described as "foaming at the mouth." The patient is usually calm in between seizures but may die during this phase from paralysis of the breathing muscles. Some human cases, as well as dog cases, can present as "dumb" rabies where there are no signs of overactivity.
- Coma and death. About ten days after the onset of symptoms, the patient slips into a coma and dies of heart failure or paralysis of the breathing muscles.

### Diagnosis

Diagnosis begins with reporting an animal bite to the doctor. The patient should describe what type of animal caused the bite and what he or she was doing when bitten. If a pet caused the bite, the patient should tell the doctor whether the cat or dog had been vaccinated against rabies. If the animal can be captured, it can be kept for ten days of observation or put to sleep at once to have its brain tissue examined.

Humans who have developed symptoms of rabies are tested by having samples of blood serum, saliva, and spinal fluid analyzed; a small piece of tissue from the nape of the neck may also be analyzed.

### Treatment

Patients who have already developed symptoms of rabies cannot be helped by immune globulin (an agent used to stimulate the immune system) or vaccine. They can be given supportive care in an intensive care unit before death.

### Prognosis

Without preventive treatment, the prognosis is death within days. Only six cases have been recorded of people surviving rabies after developing symptoms of the disease; five of the six patients had received some kind of preexposure or postexposure treatment before their symptoms appeared. The sixth survivor, a young woman from Wisconsin, is still being studied to determine what factors may have helped her survive without vaccine treatment.

### Prevention

The prevention of rabies is known as post-exposure prophylaxis or PEP. It should begin as soon as possible after a bite or other known exposure to the rabies virus. In the United States, PEP consists of one dose of rabies immune globulin, which provides temporary antibodies against the disease. It is injected in part around the site of the bite. The patient also receives five injections of rabies vaccine over a twenty-eight-day period, given in the muscle of the upper arm; the vaccine stimulates the body to produce its own antibodies against the rabies virus.

There are several measures that people can take to prevent exposure to rabies:

- Have pet cats, dogs, and ferrets vaccinated against rabies and keep their shots current.
- Avoid contact with wild or strange animals, whether alive or dead.
- Report stray animals or any animals that look sick or are acting strangely to animal control or the local police.
- Since most of the animals likely to carry rabies, except for dogs and cats, are nocturnal, any animals seen during the day and seemingly not afraid of humans should be avoided.
- Have the house batproofed according to instructions from the CDC or Bat Conservation International (BCI).
- Teach children not to pet or handle wild or unfamiliar animals.

To prevent getting rabies after exposure to a potentially rabid animal, people should:

• Wash the wound immediately with one part soap to four parts of water.

- **Negri bodies:** Round or oval bodies found within the nerve cells of animals infected by the rabies virus. They were first described by Dr. Adolchi Negri in 1903.
- **Postexposure prophylaxis (PEP):** A treatment given after exposure to the rabies virus. It consists of one dose of rabies immune globulin and five doses of rabies vaccine.
- **Prodrome:** A period before the acute phase of a disease when the patient has some characteristic warning symptoms.
- **Zoonosis:** A disease that animals can transmit to humans.

- Go to the emergency room as soon as possible after cleaning the wound.
- Contact the local animal control and public health authorities to have the animal trapped and evaluated for rabies.

People who work with animals, such as veterinarians, animal control workers, scientists who work with bats or other animals known to carry rabies, and some laboratory workers should be immunized against rabies before they are exposed, and have a booster shot every two years.

### The Future

The case of the young woman who survived rabies without vaccine has interested researchers in studying the treatment that was given to her to see whether it might benefit others. She had been placed in a coma to protect her brain as well as given a combination of drugs and vitamin replacement therapy. It is not yet known, however, whether the treatment was effective, whether she had been infected by an unusually weak strain of the rabies virus, or whether her survival resulted from both factors.

**SEE ALSO** Encephalitis; Toxoplasmosis

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## u

## **Restless Legs Syndrome**

### Definition

Restless legs syndrome is sometimes defined as a movement disorder and sometimes as a sleep disorder, because patients who suffer from it often develop insomnia. It is also known as Ekbom's syndrome, named for Karl-Axel Ekbom, a Swedish doctor who published a paper about eight patients with RLS in 1945. The oldest description of the disorder, however, was written by Thomas Willis (1621–1675), an English doctor who is considered the father of modern neurology.

### Description

Patients with RLS often find the condition difficult to describe; they may speak of it as an almost irresistible urge to move the legs, usually when they are trying to sleep. The sensations are usually only bothersome but may be painful for some patients. People with RLS may use words like "pins and needles," "like ants crawling under my skin," "stinging," "tugging," "like an electrical shock," "burning," or "creeping" to explain to the doctor what their sensations feel like.

The patient often tries to relieve the uncomfortable feelings by moving the legs or rubbing them. The person may change position in bed or get up and walk around for a few minutes. The unpleasant sensations are usually relieved while the patient is moving around but come back when the patient is trying to rest. Many people have a daily pattern to RLS, with the symptoms worsening at night and going away around daybreak.

### **Demographics**

It is thought that between 2 and 15 percent of the American population has RLS. Most people have only a mild form of the disorder, but others are severely affected.

RLS is more common in adults and often gets worse with age; however, it can occur in children and teenagers. One study of people with severe RLS found that a third of them had their first symptoms before they were twenty; by age fifty they had their sleep disrupted almost every night by the disorder.

### Also Known As

RLS, Ekbom's syndrome

#### Cause Uncertain

#### Symptoms

Burning, itching, or painful feeling in the legs relieved by movement

#### Duration

May last for years and get worse with age



*Testing for restless legs* syndrome can involve monitoring leg movements during sleep. GARO / PHOTO RESEARCHERS, INC.

RLS is thought to affect men and women equally, although some researchers report that it is more common in women.

### **Causes and Symptoms**

Restless legs syndrome has two subtypes, primary RLS, which runs in families; and secondary RLS, which may be caused by iron deficiency, pregnancy, kidney failure, or abnormalities of the nerves in the legs. Restless legs syndrome is not caused by mental disorders or by stress, but it can make them worse or be made worse by them. In some cases RLS is a side effect of certain medications—particularly cold remedies, decongestants, some types of allergy medications, and some drugs given to stop nausea and vomiting.

Several theories have been offered about the cause of RLS but none have been proven. Dr. Ekbom thought that RLS might be caused by slow circulation of the blood in the leg veins. Other suggestions include abnormal levels of neurotransmitters, which are chemicals produced by the brain that transmit impulses from one nerve cell to the next. Neurotransmitters affect muscle movement as well as moods and emotions. In 2007 a group of researchers in Iceland discovered a gene that increases a person's risk of developing RLS.

The central symptom of restless legs syndrome is uncomfortable sensations in the legs combined with an urge to move the legs in order to

### Yoga Therapy for RLS

Yoga is increasingly recommended as a treatment for restless legs syndrome (RLS), as a good overall form of exercise for maintaining flexibility of joints and muscles, and a relaxation technique to relieve stress.

Typical yoga workouts consist of a series of body postures (asanas), usually involving gentle stretching or twisting movements; the person moves gradually into the asana and holds it for a few seconds while using breathing exercises.

Asanas that increase blood circulation in the lower legs are recommended for RLS. Alice Christensen, founder of the American Yoga Association, recommends the knee squeeze and the spine twist. For the former, the person lies flat on the back with arms at the sides. Breathing in to a count of three, the person raises the right knee to the chest, wraps the arms around the knee, and holds his or her breath for a count of three while squeezing the knee against the chest. Then the person breathes out to a count of three, straightening the leg until it rests on the floor. This is repeated with the left leg.

In the spine twist, the person lies with the shoulders flat on the floor and gently rotates the spine by twisting at the waist. One knee is bent, brought up, and crossed over the body while the other leg remains on the floor. relieve the sensations. About 85 percent of patients with RLS also have periodic leg movements during sleep, and about 90 percent have trouble getting a good night's sleep. Many of these patients have problems with daytime drowsiness and depression related to loss of sleep.

### Diagnosis

There is no laboratory test for restless legs syndrome. Any imaging tests or other medical tests that are performed are done to rule out other disorders of the muscles or nervous system. The doctor usually bases the diagnosis on the patient's descriptions of how his or her legs feel and what makes them feel better. There are four criteria that define RLS, listed in 1995 by an international committee:

- The person has a strong urge to move the legs that is impossible to resist. The need to move the legs is combined with uncomfortable sensations in the legs.
- The symptoms become worse when the person is resting or sitting still.
- The symptoms are relieved very quickly when the person starts moving the legs.
- The symptoms are worse at night, especially when the person is lying down.

### Treatment

Some patients can be helped without prescription medications by cutting down on beverages containing caffeine (coffee, tea, and cola drinks), limiting their use of alcohol, and getting a healthful amount of physical exercise. Other non-drug treatments include hot baths, massaging the legs, or applying hot or cold packs.

Patients who are not helped by these treatments may be given one or more types of drugs to relieve their symptoms. One type of drug works

**Neurology:** The branch of medicine that studies and treats disorders of the nervous system.

**Neurotransmitters:** Chemicals produced in the brain that transmit nerve impulses to other nerve cells and eventually to muscles.

**Primary disease:** A disease that develops by itself and is not caused by a previous disease or injury.

**Secondary disease:** A disease that is caused by another disease or condition or by an injury.

by changing the levels of neurotransmitters in the brain. Other groups of drugs used to treat RLS include tranquilizers, painkillers, and drugs used to treat epilepsy. The doctor may have to try more than one type of medication before finding one that will work for the specific patient.

Patients who have RLS because of an iron deficiency in their blood can be given iron supplements.

### Prognosis

The prognosis of the disorder depends on the person's age and whether he or she has the primary form or the secondary form of RLS. Older people are less likely to benefit from treatment. Women who develop RLS during pregnancy often feel better after the baby is born.

### Prevention

There is no known way to prevent RLS because the causes of the disorder are not fully understood.

### **The Future**

Genetic research may help scientists to better understand the causes of RLS and then develop a treatment that will work for everyone with the disorder.

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## **u** Reye Syndrome

#### Definition

Reye syndrome is a rare but acute illness characterized by brain damage and liver failure following a viral infection, most commonly an infection of the upper respiratory tract, influenza, chickenpox, or a stomach disorder. It occurs most often in children but has been reported in adults. Adults, however, usually recover completely, whereas children who develop the syndrome have a high risk of lasting brain damage or death.

#### Description

Reye syndrome is a life-threatening condition marked by swelling of the brain and a buildup of fatty deposits in the liver. What triggers the syndrome is not fully understood. It is sometimes described as a two-phase illness because it is almost always preceded by a viral infection.

#### Demographics

Reye syndrome is most likely to occur in children between the ages of five and fifteen; it appears to be more common in Caucasians in the United States than in members of other racial groups. Reye syndrome is reportedly most likely to occur in January, February, or March, during

#### Also Known As

Reye-Johnson syndrome, acute noninflammatory encephalopathy

#### Cause

Unknown; may be triggered by viral infections

#### Symptoms

Vomiting, sleepiness, paralysis, seizures, loss of consciousness, death

#### Duration

Can be fatal within a few days



Liver cells from a child who died of Reye syndrome. Pale spots in the cells are accumulations of fat. SCIENCE SOURCE.

influenza season. Epidemics of chickenpox or flu were sometimes followed by a rise in the number of reported cases of Reye syndrome.

In general, however, the disorder is much less common in the United States in the early 2000s than it was in 1980, when 555 cases were reported to the Centers for Disease Control and Prevention (CDC). In 1999, the CDC reported that the annual number of cases had declined rapidly, to an average of thirty-seven each year between 1987 and 1993. Since 1994, only about two cases per year have been reported in the United States.

### **Causes and Symptoms**

The cause of Reye syndrome is still unknown. It is known that the condition is not contagious. In addition, the reasons for the decline in the number of cases since 1980 are not clear. Some doctors think that there are fewer cases of Reye syndrome at present because parents have heeded the warnings required by law since 1986 on all packages containing aspirin. Others wonder whether Reye syndrome is triggered by a specific strain of chickenpox or influenza virus rather than by aspirin because the viruses that cause these diseases are constantly mutating, or changing. In 2004 some doctors in Florida argued that the syndrome was caused by "a viral mutation which spontaneously disappeared" or by inherited metabolic disorders that were not recognized in the 1950s and 1960s. In addition, several doctors in Japan have reported cases of Reye syndrome in children who had not been given aspirin.

### The Discovery of Reye Syndrome

Ralph Douglas Reye (1912–1977; name pronounced like rye) was a pathologist who worked at the Royal Alexandria Hospital for Children in Sydney, Australia, in the early 1950s. In 1951 a pediatrician brought him a case involving a ten-month-old boy who died after thirty hours of vomiting. The pediatrician thought it might have been an infection, but Dr. Reye found liver and brain damage of a type that he had never seen before when he performed an autopsy. Over the next decade Dr. Reve identified twenty more cases of this strange syndrome. In 1963 he published an article in a prestigious British medical journal about his cases. A link with aspirin was not made until two younger doctors who were working with Dr. Reye interviewed the families of the twenty-one children he had studied and found that eleven of the children had taken aspirin during their illness.

Dr. Reye's work was confirmed by Dr. George Johnson, an American doctor who reported on the deaths of 16 school-age children in North Carolina in the mid-1960s who had the same kind of liver and brain damage as the children that Dr. Reye had studied. By 1986, the Food and Drug Administration (FDA) required American drug manufacturers to place a warning about Reye syndrome on all bottles of aspirin produced in the United States and to remove aspirin from most over-the-counter products intended for children. The symptoms of Reye syndrome are linked to the buildup of fatty deposits in the liver and increased pressure on the brain. The liver is not able to get rid of ammonia, a substance that is produced during the digestion of proteins in food. The ammonia builds up in the blood, a condition called hyperammonemia. This condition in turn can be associated with brain damage.

The symptoms of Reye syndrome are sometimes classified into early and later symptoms. It is important to note, however, that not all children with Reye syndrome have all these symptoms or develop them in the same order. Another important fact to keep in mind is that fever is *not* usually present in Reye syndrome.

Early symptoms of the disorder include:

- Persistent or continuous vomiting
- In small children, diarrhea and rapid breathing
- Unusual sleepiness and loss of energy (This symptom usually appears about twenty-four hours after the beginning of the vomiting.)

The second stage of Reye syndrome is marked by personality changes and changes in level of consciousness:

- Irritability and aggressive behavior
- Mental confusion
- Weakness or paralysis of the arms and legs
- Seizures
- Loss of consciousness and coma

### Diagnosis

There is no single test that is specific for Reye syndrome. It is diagnosed primarily by excluding other possible causes of the child's symptoms. Because Reye syndrome is a serious illness, however, the doctor must consider it as a possible diagnosis in any child with a history of a recent viral illness combined with severe vomiting and changes in the level of consciousness.

The doctor will give the child the following types of tests:

- Blood and urine tests. These can help the doctor rule out the possibility of accidental poisoning or exposure to a toxic substance.
- Spinal tap (also called a lumbar puncture) to take a sample of cerebrospinal fluid (CSF). CSF is the fluid that surrounds the brain and the spinal cord. Analyzing the sample can help to determine whether the illness is caused by a brain infection rather than Reye syndrome.
- Liver biopsy. This test is performed by inserting a needle into the right side of the child's abdomen and removing a small sample of liver tissue.
- Computed tomography (CT) or magnetic resonance imaging (MRI) scans. These tests provide images of the brain that can help the doctor rule out tumors, head injuries, or other causes of the child's changes in behavior or level of alertness.

### Treatment

Treatment of a child thought to have Reye syndrome must begin as soon as possible to lower the risk of death or lasting damage to the nervous system. Children diagnosed with Reye syndrome require treatment in the intensive care unit (ICU) of a hospital. There is no specific drug or other type of treatment that is unique to Reye syndrome. The child is carefully monitored for changes in vital signs (blood pressure, pulse or heart rate, breathing rate, and temperature) and symptoms and may be given one or more of the following:

- Intravenous fluids containing glucose (sugar) to nourish the body as well as replace lost fluids
- Small amounts of insulin
- Steroid medications to reduce swelling of the brain tissue
- Diuretics, drugs that speed up urine output
- If necessary, a respirator to help the child breathe

### Prognosis

Most children treated early make a full recovery, but about 20 percent of children with Reye syndrome will die even with prompt treatment. About 10 percent of survivors suffer some kind of permanent damage

**Ammonia:** A chemical produced during the breakdown of protein in the body. It is usually converted in the liver to another chemical called urea and then discharged from the body in the urine.

**Diuretics:** Medications that speed up the production of urine. **Hyperammonemia:** Overly high levels of ammonia in the blood; it often indicates liver damage.

Vital signs: Measurements taken to evaluate basic body functions. They are temperature, pulse rate, blood pressure, and breathing rate.

to the nervous system; the extent of the damage depends on the ammonia level in the blood. If treatment is delayed, the death rate can rise substantially. Children under the age of five are at greatest risk of severe brain damage or death.

### Prevention

The best way to prevent Reye syndrome is to have children vaccinated against influenza and chickenpox and to avoid giving aspirin to children and teenagers under the age of eighteen when they get a cold or flu-like illness.

#### The Future

Further research may shed light on either the cause of Reye syndrome or the reasons for the drop in the number of cases reported since the 1990s.

SEE ALSO Chickenpox; Influenza

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## **Rheumatic Fever**

### Definition

Rheumatic fever is caused by an inflammatory reaction that occurs after an infection caused by certain Group A streptococci, sphere-shaped bacteria that grow in long chains. Strep throat and scarlet fever are the illnesses most likely to lead to rheumatic fever if they are not properly treated.

### Description

Rheumatic fever has been known for centuries; its symptoms were described in the seventeenth century by Thomas Sydenham (1624–1689), an English doctor, although the connection between rheumatic fever and bacterial infections was not yet known. In 1880 doctors discovered that a sore throat could lead to rheumatic fever; in the early 1900s they found that there was also a link between scarlet fever and rheumatic fever. The diagnostic criteria for rheumatic fever, still used in the early 2000s, were first identified by a doctor named T. Duckett Jones in 1944. There were no effective preventative treatments for rheumatic fever, however, until the late 1940s, when antibiotics proved to be effective treatments for both strep throat and scarlet fever.

Although fever is one of the symptoms of rheumatic fever, the disease is better understood as an inflammatory disorder that affects several body systems, particularly the skin, the tissues under the skin, the central nervous system, the joints and muscles, and the circulatory system. Multiple episodes of rheumatic fever can cause long-term damage to the heart and its valves that gets worse over time. This complication is known as rheumatic heart disease. Rheumatic fever can recur even after antibiotic treatment for the acute phase of the disease.

Rheumatic fever develops in about 3 percent of people who have either untreated strep throat or scarlet fever about twenty days after the strep infection. In about 30 percent of cases, the patient was not treated for the strep infection because he or she had no noticeable symptoms. A common pattern for rheumatic fever is pain and swelling in the joints between two and six weeks after the strep infection, along with fever and a generally unwell feeling. The patient may also develop lumps of tissue

#### Cause

Response to a streptococcal bacterial infection

#### Symptoms

Painful joints, rash, skin nodules, abdominal pain, fever, nosebleeds, heart problems

#### Duration

Lifelong complications if untreated

#### **Rheumatic Fever**

Image of inflamed heart muscle (myocarditis) caused by rheumatic fever. The spaces between the fibers (white and blue) show fluid accumulation. ASTRID & HANNS-FRIEDER MICHLER / PHOTO RESEARCHERS, INC



known as nodules beneath the skin of the wrists, elbows, and knees, and a rash on the chest and upper arms. Another symptom is chorea, a movement disorder that affects the muscles of the patient's face and upper arms. The symptoms of the initial attack of rheumatic fever go away in about six weeks for 75 percent of patients; the remainder may take as long as twelve weeks to recover.

People who have had an acute attack of rheumatic fever are more likely to have the disease flare up again if they get another strep infection. These recurrences can lead to severe heart valve damage and can predispose people to bacterial endocarditis, a condition that develops when bacteria form colonies on the tissues of the heart valves. Pieces of damaged tissue can break off from the valves and travel through the bloodstream, eventually blocking major blood vessels and causing stroke or heart failure.

#### Demographics

Rheumatic fever used to be much more common before the introduction of antibiotics in the 1940s than it is in the twenty-first century. According to the American Heart Association, about 15,000 people in the United States died each year of rheumatic fever and its complications in the 1950s. In 2004 there were 3,248 deaths resulting from rheumatic fever, 2,226 in women and 1,022 in men. The disease is rare in the

### A Murder Mystery Solved

For over 200 years, music scholars and biographers wrestled with a conspiracy theory about the sudden death of Wolfgang Amadeus Mozart (1756–1791) just a few weeks before the famous composer's thirty-sixth birthday. Some thought that Mozart might have been poisoned by Antonio Salieri, a rival musician who was jealous of Mozart's talents. In 2000, however, a professor of medicine at the University of California, Davis, reopened the case by looking at the composer's symptoms as reported by his family and doctors at the time. She concluded that Mozart died of complications from rheumatic fever, not murder.

The professor noted that Mozart had experienced several episodes of rheumatic fever earlier in his life, which almost certainly damaged his heart. In late November 1791 he fell ill suddenly with fever, headaches, a skin rash, and painful swelling of the joints in his arms and legs. In the second week of his illness, his body swelled with fluid to the point where his clothes did not fit and he could not sit up in bed without help—indications of congestive heart failure. Mozart then lost consciousness and



*Statue of Wolfgang Amadeus Mozart in Vienna, Austria.* SHUTTERSTOCK.

died on December 5, 1791, after just fifteen days of illness.

United States and other developed countries as of 2008, but is still a major killer in less developed parts of the world. A recent report indicates that 95 percent of cases of rheumatic fever worldwide occur in the poorer nations, particularly those with tropical climates.

People of any age can get rheumatic fever after a strep infection; however, most cases in North America are diagnosed in children between the ages of five and fifteen. As far as is known, rheumatic fever in the United States affects both sexes and all races equally, although people of any race living in crowded conditions are at increased risk. In terms of symptoms, girls are more likely than boys to develop chorea and heart valve problems. The disease is most likely to occur in the winter—the peak season for strep throat and scarlet fever.

### **Causes and Symptoms**

As of 2008, rheumatic fever is thought to result from an abnormal response of the immune system to certain Group A streptococci. There is no evidence that infections involving other types of bacteria can lead to rheumatic fever. Some doctors also think that there may be genetic factors that make some people more susceptible to rheumatic fever.

The most significant symptoms of rheumatic fever include:

- Fever
- Chorea: Jerky involuntary movements of the arms and facial muscles
- Painful swollen joints
- Chest pain
- Fatigue
- A faint pink rash on the upper body, which may have a ringshaped or snake-like pattern
- Nodules on the wrists, elbows, and knees

Other symptoms may include shortness of breath, pain in the abdomen, and nosebleeds.

#### Diagnosis

The diagnosis is based on a combination of the patient's history (particularly a strep infection within the previous two months), the patient's descriptions of his or her symptoms, and the results of laboratory tests used to evaluate the symptoms. There is no single test that can diagnose rheumatic fever. The present guidelines for diagnosis state that the patient must meet either two major diagnostic criteria or one major criterion and two minor ones. The major criteria for diagnosing rheumatic fever are:

- Evidence of carditis (inflammation of the heart). The signs of carditis include heart murmurs, congestive heart failure, or shortness of breath and chest pain.
- Arthritis in several joints.
- Nodules under the skin.
- Chorea.
- Skin rash.

The minor criteria include fever; joint pain; abnormal findings on an electrocardiogram (ECG); blood test results that indicate a recent strep infection; and a history of previous episodes of rheumatic fever.

Carditis: Inflammation of the heart.

**Chorea:** A movement disorder characterized by brief involuntary movements of the muscles.

**Diuretics:** A group of medications given to increase urine output.

**Endocarditis:** Inflammation of the lining of the heart and its valves.

**Group A streptococcus:** A sphere-shaped bacterium that grows in long chains and causes strep throat as well as scarlet fever.

**Nodule:** The medical term for a small rounded lump of tissue.

#### Treatment

Acute rheumatic fever is treated with antibiotics to kill the bacteria and to prevent future flareups of rheumatic fever. Penicillin is the most common antibiotic used. Patients are typically maintained on daily (by mouth) or monthly (by injection) doses of antibiotics for three to five years after the first episode of rheumatic fever; in some cases, they may be given maintenance doses of penicillin for life, particularly if they have suffered damage to their heart valves or are at high risk of repeated exposure to streptococcal infections.

Patients are also given over-the counter pain relievers like aspirin or Tylenol for joint pain and fever. Those with severe carditis are given corticosteroid medications to reduce the inflammation in the heart tissue. Patients who have developed congestive heart failure are treated with diuretics and a heart drug called digitalis.

### Prognosis

The prognosis of rheumatic fever depends on whether the patient developed carditis and if so, how severely the heart was damaged. Bacterial endocarditis can lead to eventual death from stroke or heart failure.

### Prevention

Rheumatic fever can be prevented by prompt diagnosis and treatment of scarlet fever or strep throat. The most effective form of prevention is giving antibiotics within nine days of the first symptoms of a Group A streptococcal infection.

### The Future

At present the National Institutes of Health (NIH) is conducting clinical studies of the rates of rheumatic heart disease in Central America and parts of Asia, as well as using DNA typing to look for possible genetic factors in rheumatic fever.

SEE ALSO Heart failure; Scarlet fever; Sore throat; Strep throat

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## Rheumatoid Arthritis

### Definition

Rheumatoid arthritis, or RA, is an autoimmune disorder characterized by inflammation of the joints, resulting in pain, swelling, and gradual loss of

function. The joints most commonly affected are the hands, wrists, elbows, knees, ankles, and feet; however, other joints may be involved as well.

### Description

RA develops gradually in about 90 percent of persons diagnosed with it. The patient notices that the affected joints do not move as freely as they once did, and there may be pain, redness, and swelling as well. Patients whose hands and wrists are affected may notice nodules (small lumps) underneath the skin of the joints. Nodules also develop on the elbows of about 20 percent of patients with RA. In most cases, the joints on both sides of the body are affected; this symmetrical pattern is one significant difference between rheumatoid arthritis and osteoarthritis. The intensity of the pain varies from person to person, but it is common for patients with RA to feel that their joints are stiffest in the morning, with some improvement later in the day.

In addition to pain and stiffness in the joints, patients with RA frequently suffer from aches in the muscles, fever and flu-like symptoms, loss of appetite, cold or sweaty hands and feet, unintended weight loss, and general lack of energy.

The symptoms of RA are not constant but tend to come and go. A period of freedom from symptoms is called a remission, and the sudden return or worsening of symptoms is called a flare. Over time, however, the symptoms of joint damage become worse. One reason for diagnosing RA as quickly as possible is to slow or prevent further damage to the joints.

RA can affect other body systems, including the skin, eyes, bones, lungs, and nervous system. Patients may experience dry eyes; tingling or numbness in the hands or feet; anemia; osteoporosis; increased vulnerability to infections; and an increased risk of lymphoma (cancer of the lymphatic system).

### **Demographics**

RA is one of the most common inflammatory joint diseases, affecting at least 2.1 million people in the United States. It strikes about three people in every 10,000 worldwide, although some countries and ethnic groups have higher rates than others. Some Native American tribes have rates of RA as high as 5 to 6 percent, while natives of the Caribbean have a lower than average rate. The reason for these differences is not yet known.

RA is primarily a disease of adults; the most common age at onset is thirty-five to fifty. Children, however, can also get rheumatoid arthritis, Also Known As RA

#### Cause

Unknown; possibly partly genetic

#### Symptoms

Painful and stiff joints, nodules beneath the skin of joints, fatigue, fever

**Duration** Years

#### **Rheumatoid Arthritis**

Deformity in the toes of a patient with rheumatoid arthritis. DR. P. MARAZZI / PHOTO RESEARCHERS, INC.



as can elderly adults. As with lupus and other autoimmune disorders, women are three times as likely as men to develop RA. Men with RA, however, are more likely to have a more severe form of RA and to die earlier than women with the disease.

Although rheumatoid arthritis is not considered a genetic disease in the strict sense, it does appear to run in some families. A parent, child, or sibling of a person with RA has a 3 percent greater risk of developing the disease themselves than someone in the general population.

### **Causes and Symptoms**

The cause of RA is not known. Some researchers think that it may be caused by a bacterium or virus because of the inflammation that weakens the patient's joints. Others think that hormones may be involved because RA affects women significantly more often than men. Still others think that genetic factors are also involved because at least four separate genes known to regulate the immune system's responses have been associated with an increased risk of RA. Some of these genetic markers, however, are also found in persons who do not have the disease. What seems clear is that there is no single gene that determines whether a person will develop RA.

What happens in rheumatoid arthritis is that the synovium, a type of tissue that lines the joints in the body, becomes inflamed, swells, and starts to damage the bones and cartilage in the joint. The inflammation can spread to other tissues near the joint, such as the tendons, ligaments, blood vessels, and nerve endings. As the tissues inside the joint are damaged by the inflamed synovium, the person experiences the destruction as stiffness and pain when using the joint. As the tissues surrounding the joint are affected by the disease, the effects of the inflammation may spread to the blood vessels, the lungs, or other organ systems. Women with RA may also develop Sjögren syndrome, a disorder characterized by dryness of the eyes and mouth.

The symptoms of RA have already been described.

### Diagnosis

Rheumatoid arthritis is not always easy to diagnose just from the patient's history because the symptoms may develop slowly over time and there are other disorders that have some of the same symptoms. In particular the doctor will need to rule out Lyme disease, osteoarthritis, and lupus. There is no single test that can be used to confirm the diagnosis of RA. The diagnosis is based on a combination of blood tests, imaging studies of the patient's joints, and a history of the patient's symptoms. The most widely accepted blood test is called the anti-CCP test; it measures the presence of an autoantibody in the patient's blood. Another autoantibody that is measured for diagnostic purposes is rheumatoid factor, or RF.

In 1987 the American College of Rheumatology drew up a list of diagnostic criteria that doctors can use to help with the diagnosis. A patient who meets four of the seven criteria is considered to have RA:

- Morning stiffness lasting longer than an hour most mornings for at least six weeks
- Arthritis in the hand joints for six weeks or longer
- Arthritis in three or more of fourteen other joints for at least six weeks
- Symmetrical arthritis for at least six weeks
- Rheumatoid factor (RF) in the blood higher than that found in 95 percent of the population
- Nodules beneath the skin
- X-ray evidence of joint damage

### Treatment

There is no cure for rheumatoid arthritis. Therapy is focused on relieving the patient's pain, slowing or preventing further damage to the joints, reducing inflammation, and improving the person's ability to function. It is critical to begin treatment as soon as possible after diagnosis. In the recent past, doctors concentrated on relieving pain and waiting for symptoms to get worse before prescribing stronger pain relievers. It is now known that joint destruction can begin within one to two years of the earliest symptoms of the disease, so that slowing down its progress is important.

The mainstay of treatment for RA is medications. These typically include several different types of drugs:

- Drugs to relieve pain and reduce inflammation. These include steroid medications and NSAIDs like aspirin and ibuprofen.
- Disease-modifying anti-rheumatic drugs (DMARDs). DMARDs are prescribed to slow down the destruction of the joint tissues. They include drugs like methotrexate, injectable gold, and penicillamine.
- Biologic response modifiers. Also known as biologics, these drugs work by blocking certain proteins that contribute to inflammation. They include drugs like Enbrel and Humira.

Most patients benefit from a combination of drugs rather than just one. Drug therapy for RA is highly individualized, and the doctor may have to try more than one drug in each group to see which works best for the patient. Combining the DMARD methotrexate with one of the biologics appears to work well for patients with moderate or severe RA.

In some cases the doctor may recommend surgery to reduce inflammation or improve range of motion in the damaged joints. The types of surgery most commonly performed to treat RA include total joint replacement; repair or reconstruction of damaged tendons and ligaments; and surgical removal of inflamed synovium.

Patients with RA are also given physical therapy or occupational therapy and an exercise program to keep their joints as flexible as possible as long as possible. Although exercise must be balanced with rest, it can help people to sleep better, improve their mood, and lose weight if needed as well as keeping joints and muscles strong.

Doctors also recommend stress reduction programs for patients with RA. Although emotional stress does not cause the disease, the anger and fear that many patients experience when they are diagnosed with RA can make the pain worse and make it more difficult to cope with the disease.

- **Autoantibody:** A type of protein made by a person's immune system that attacks the body's own tissues.
- Flare: A return or worsening of the symptoms of RA.
- **Remission:** A period of freedom from the symptoms of RA.
- Rheumatoid factor (RF): An antibody that attacks the body's own tissues that is found in some

patients with RA and is measured as part of the diagnostic process.

- **Rheumatologist:** A doctor who specializes in diagnosing and treating arthritis and other diseases of the muscles and joints.
- **Synovium:** A type of tissue lining the joints that ordinarily secretes a fluid that lubricates the joints.

Relaxation techniques, meditation, and support groups are all beneficial ways to lower a patient's stress level.

### Prognosis

RA is a disease that involves significant loss of function and shortened life spans for most patients diagnosed with it. Only a small minority—about 5 percent—of patients recover spontaneously. About a third of patients are forced to stop working within five years of being diagnosed; after ten years, about 50 percent of patients have some limitations on their activity. People with a family history of RA have worse prognoses than those without; men generally have worse prognoses than women; and those who are elderly at the time of diagnosis have worse prognoses than younger patients. Researchers estimate that patients with RA have their life expectancy shortened by five to ten years on average.

### Prevention

There is no known way to prevent rheumatoid arthritis; however, early diagnosis and prompt treatment can slow the progression of the disease and reduce the patient's risk of complete disability.

### The Future

Researchers are presently looking at new treatments for rheumatoid arthritis as well as trying to gain a better understanding of its causes. There are over 600 clinical studies under way as of 2008 looking at new biologic drugs as well as various combinations of DMARDs and biologics in treating the disease. The National Institutes of Health and the Arthritis Foundation have set up ten research centers around the United States that are collecting genetic materials from families in which two or more siblings have developed rheumatoid arthritis. Other researchers are studying the role of hormones in RA to discover why the disease affects women more frequently but men more severely.

Another question researchers are investigating is why the number of new cases of RA has started to drop in the United States in recent years. Although the decline is not a sharp one, it may shed new light on the cause or causes of RA.

**SEE ALSO** Fibromyalgia; Lupus; Lymphoma; Osteoarthritis; Osteoporosis; Sjögren syndrome

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## Rubella

### Definition

Rubella is a mild viral illness characterized by a rash that in some ways resembles the reddish rash of measles. However, the two diseases should not be confused. Rubella is sometimes called German measles because it was first described by German doctors in the eighteenth century.

### Description

Rubella is a contagious illness transmitted from person to person by droplets in the coughing or sneezing of an infected person. It is not as contagious as measles, however. The disease is mild and may sometimes not be noticed, particularly in children, who recover more quickly than adults. In most people, rubella runs its course in two to five days.

Rubella has a fairly long incubation period of two to three weeks. The most important complication of rubella is congenital rubella syndrome (CRS), a condition that can develop in some babies born to mothers who had rubella during the first three months of pregnancy. About 20 percent of these pregnancies end in miscarriage. Babies with CRS may be born with incurable birth defects such as blindness; mental retardation; abnormalities of the heart, eyes, or brain; low birth weight; and disorders of the liver or spleen. In later childhood, they are at increased risk of glaucoma, movement disorders, autoimmune disorders, and mental disorders such as schizophrenia.

### Demographics

Rubella is much less common in the United States than it was before the development of the measles-mumps-rubella (MMR) vaccine in 1969. In 1963 and 1964, there was a rubella epidemic in the United States and twelve million people were infected with the disease, including thousands of pregnant women. According to the Centers for Disease Control and Prevention (CDC), 11,000 babies died before birth and another 20,000 were born with CRS. After the vaccine became available, the number of rubella cases dropped rapidly, to about 1,000 per year by the early 2000s. There are only about ten cases of CRS each year in the

Also Known As German measles, threeday measles

### Cause

Virus

#### Symptoms

Headache, low-grade fever, runny nose, bloodshot eyes, reddish skin rash, muscle or joint pain

Duration

Two to five days

#### Rubella

Rubella rash on a woman's arm. DR. P. MARAZZI / PHOTO RESEARCHERS, INC.



United States at present, most of them involving Hispanic mothers who were not vaccinated.

Before 1969, rubella was most common among children between the ages of five and fourteen years. In recent years most people infected in the United States are older teenagers or young adults. Males and females are equally likely to get rubella.

### **Causes and Symptoms**

The cause of rubella is a virus that typically enters the body through the throat or the nasal passages. People can also get the disease by touching a soiled tissue or handkerchief that was used by an infected person. The virus reproduces itself in the moist tissues that line the nose and throat. It then moves into the lymph nodes and from there into the central nervous system and the bloodstream, usually five to seven days after infection. In pregnant women, the virus can infect the baby through the mother's blood.

The first symptoms of rubella are a generally unwell feeling and loss of appetite. These are followed by a reddish or pinkish rash that begins on the face and spreads downward over the rest of the body; swollen glands in the neck and behind the ears; a low-grade fever of 101°F (38.3°C); joint pains; headache; a runny nose; and reddened eyes. The rash may itch but usually goes away in a few days without staining the skin like the characteristic rash of measles. A person with rubella is contagious from one week before the onset of the rash until about a week after the rash disappears, but is highest just before and on the day that symptoms appear. Children rarely have any complications with rubella but older teenagers or adults may develop joint pains similar to those of arthritis. In most adults, these joint pains go away in several months to a year.

### Diagnosis

In some cases rubella can be diagnosed by the doctor on the basis of the physical symptoms. Some doctors, however, will order a virus culture made from a throat swab or a blood test to confirm the diagnosis.

### Treatment

Rubella is a mild disease that does not require special treatment in most cases. Patients can rest in bed and take acetaminophen or another non-aspirin pain reliever to bring down the fever. They should, however, stay away from other people and tell friends and family members (especially pregnant women) that they may have been exposed to rubella.

### Prognosis

In a very few cases, people may develop an ear infection or encephalitis (viral infection of the brain) as complications of rubella, but these are very rare.

### A Rubella Tragedy

Gene Tierney (1920–1991) was a glamorous actress who starred in a number of stage plays and films in the 1940s and 1950s. Married to a fashion designer named Oleg Cassini, Tierney became pregnant with her first child, a daughter, in early 1943. In June, Tierney caught German measles while making an appearance at a Hollywood club for people in the military. She later found out that she had been infected by a woman in the Marine Corps who knew she had rubella but snuck out of her barracks under quarantine to meet Tierney and ask for her autograph.

Tierney's daughter Daria was born prematurely in October 1943, weighing little more than three pounds. The little girl was also deaf, partially blind, and severely mentally retarded. She eventually required full-time institutional care. Tierney later wrote in her autobiography, "After that I didn't care whether ever again I was anyone's favorite actress." In 1962, the famous detective writer Agatha Christie (1890– 1976) published a mystery titled *The Mirror Crack'd from Side to Side*. Its plot line was based on Tierney's tragedy. The major difference is that in Christie's story, the woman who infected the actress with rubella is poisoned in revenge.

Most people recover completely in a week without lasting effects.

The prognosis for children affected by congenital rubella syndrome is poor; many die at an early age from heart defects and other abnormalities. Those who survive childhood usually require specialized care for blindness, deafness, or mental retardation.

### Prevention

A person who has had rubella is protected for life against becoming infected with the disease again. For children who have not had the

**Congenital rubella syndrome (CRS):** A group of birth defects that may affect a baby born to a mother who had rubella during the first three months of pregnancy. **Lymph node:** Part of the lymphatic system, the lymph nodes trap foreign particles and are important to defend the body from disease.

disease, it can be easily prevented by two doses of the combined measlesmumps-rubella (MMR) vaccine, usually given between twelve and fifteen months of age for the first shot, and between four and six years of age (before school entry) for the second vaccination.

Women of childbearing age who are not sure whether they received the MMR vaccine as children or are otherwise immune to the virus can have a blood test to find out whether antibodies against the virus are present. If not, they can be immunized as long as they postpone getting pregnant for twenty-eight days after the vaccination. Women should not be vaccinated against rubella at any time during pregnancy, or if they have an immune system weakened by cancer chemotherapy, radiation treatment, or HIV infection. People who are moderately or severely ill with any disease or disorder should consult their doctor before being vaccinated.

#### The Future

Rubella is likely to become an increasingly uncommon disease in the United States. Nonetheless, children should continue to be immunized against it, as periodic outbreaks occur from time to time when Americans travel to countries where immunization against the disease is not routine. Only about half of the world's population had been immunized against rubella as of 2008. Most recent outbreaks in the United States have been among Hispanics, many of whom were born in countries without rubella vaccination programs.

SEE ALSO Developmental disability; Glaucoma; Measles

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# S



## Genetic

Infection

Injury

Multiple

Other

Unknown

#### **Also Known As** Scarlatina

#### Cause

Bacterium (Group A streptococcus)

### Symptoms

Fever, sore throat, vomiting, red rash with sandpapery feel, strawberry tongue, skin peeling

#### Duration

About ten days; rash and skin peeling may take two to three weeks to clear



## Scarlet Fever

### Definition

Scarlet fever is an infectious disease caused by certain strains of the bacteria (Group A streptococcus) that cause strep throat. It is characterized by a high fever, a red rash with a sandpapery texture that covers most of the body, and a so-called strawberry tongue.

### Description

Scarlet fever is an infectious disease of childhood that can occur together with strep throat. It is spread by fluids from the throat or nose of an infected person or by contact with a drinking glass, soiled tissue, or other object touched by an infected person. The incubation period is short; children can feel sick within twelve hours of being infected, although the usual incubation period is one to two days. Children are contagious from the moment the acute symptoms appear until the rash completely fades.

The first phase of scarlet fever is a prodrome, or warning period marked by a general feeling of tiredness, headache, sore throat, and fever. In the acute phase of scarlet fever, which begins about two days after the child first feels sick, the characteristic rash appears, first on the neck and then spreading downward to the rest of the body. The child often has flushed or reddened cheeks with the area around the mouth unusually



Young man with a rash from scarlet fever. © SALLY AND RICHARD GREENHILL/ALAMY.

pale. The rash begins to fade in about three or four days, followed by the peeling of the skin on the face, palms of the hands, and fingers.

Scarlet fever tends to be a milder disease than it was before the use of antibiotics to treat it in the 1950s. It can, however, lead to rheumatic fever, inflammation of the kidneys, or certain types of psychiatric disorders (grouped together as PANDAS disorders) if it is not promptly treated.

### **Demographics**

Scarlet fever is most likely to affect children between the ages of five and fifteen years; it is unusual in people over eighteen. It affects boys and girls equally, and appears to be equally common in all races and ethnic groups. In the United States it is most likely to occur in winter and spring.

Scarlet fever is thought to occur in 1 percent of the school-age population, or about 10 percent of children diagnosed with strep throat.

### **Causes and Symptoms**

Scarlet fever is caused by a Group A streptococcus that normally lives in the upper respiratory tract. In many cases the disease follows an episode of strep throat. As the bacteria grow in the tissues of the nose and throat, they secrete a toxin that is responsible for the rash associated with scarlet fever.

During the prodrome, the child may have a fever between 101 and  $104^{\circ}F$  (38.3°C and 40°C), along with:

- Sore throat
- Headache
- Vomiting
- Abdominal cramps
- Enlarged glands in the neck that are sore to the touch
- Aching muscles

### The World's Fastest Woman

Scarlet fever is rarely considered a serious threat to children's health, but in the 1940s it was still a common cause of disabilities, including complications affecting the joints and muscles. Wilma Rudolph (1940–1994), the first American woman to win three gold medals in track and field in a single Olympic Games, could not even walk without braces until she was twelve years old. Rudolph suffered from polio and pneumonia as well as scarlet fever in her early years. She was treated at home by her mother; the future champion later remembered that during her youth, "My mother used to have all these home remedies she would make herself, and I lived on them."

After starting high school, Rudolph became a basketball star and earned a position on the U.S. women's track and field team for the 1956 Olympics. She was only sixteen years old at the time. In 1960 she won three gold medals at the Rome Olympics, running the 100-meter dash in only eleven seconds and being hailed as "the fastest woman in history." In 1963 Rudolph completed her college education at Tennessee State University and became an elementary school teacher.

Rudolph did not forget her early struggles with illness and the importance of her mother's



Wilma Rudolph at the 1960 Summer Olympics. ALLSPORT.

encouragement. In 1981 she started a foundation to help other young athletes by teaching them that success is possible even in the face of sickness and other difficulties. Her foundation provides free coaching in the young people's chosen sports. She died of brain and throat cancer in 1994, but her work lives on in the accomplishments of the athletes she has inspired and helped.

When the rash appears, it is sometimes described as looking like a bad sunburn. The sandpapery texture of the rash is its most important feature. It may itch and will typically turn white when pressed with a finger. Other symptoms of this phase of the illness include:

- Chills.
- Fast heartbeat.
- Small pinpoint spots on the inside of the mouth caused by tiny broken blood vessels. These are sometimes called Forchheimer spots.

- Appearance of the tongue changes from a whitish-looking coating over red and swollen taste buds to a completely red and swollen tongue.
- Appearance of Pastia's lines. These are lines of bright red color that appear in the folds of the body—the armpits, elbows, neck, knees, and groin area.
- Peeling of the rash about four or five days after its appearance.
- White or yellowish patches on the tonsils.

### Diagnosis

The diagnosis of scarlet fever is usually based on a combination of the patient's history—particularly recent exposure to other children with strep throat or scarlet fever—and the doctor's examination of the patient's throat, neck glands, and skin. The doctor may also take a sample of fluid from the child's throat on a cotton swab. The fluid can be sent to a laboratory for a throat culture, which is an accurate test that takes two days. The doctor may also perform what is called a rapid strep test. It can detect the streptococci in a few minutes in the doctor's office. The rapid strep test, however, is not as accurate as a throat culture.

A newer test for scarlet fever is called a rapid DNA test, which is as accurate as a throat culture but takes less than a day to give results.

### Treatment

Patients with scarlet fever are treated with a course of antibiotics, most commonly penicillin or a similar drug known to be effective in treating strep throat. The child will usually not be contagious within a day or two of starting treatment and can return to school once the fever goes down. It is important, however, for the child to take the full course of antibiotics (usually ten days), even though he or she may start to feel better in a few days. Peeling of the skin may continue for another two or three weeks, but it does not mean that the child is still contagious.

The doctor may recommend a soft or liquid diet if the child's throat is very sore and he or she finds it hard to eat. Warm soups and teas, milkshakes, ice cream, and soft drinks are all good choices. It is important for the child to drink plenty of fluids to prevent dehydration.

In a few cases, the doctor may recommend a tonsillectomy to prevent recurrence of streptococcal infections.

- **Forchheimer spots:** Tiny reddish spots that appear inside the mouth of a patient with scarlet fever. They are named for Frederick Forchheimer (1853–1913), an American doctor.
- **Group A streptococcus:** A sphere-shaped bacterium that grows in long chains and causes strep throat as well as scarlet fever.
- **PANDAS disorders:** A group of disorders with psychiatric symptoms that develop in some children after strep throat or scarlet fever. The acronym stands for Pediatric Autoimmune

Neuropsychiatric Disorders Associated with Streptococcal infections.

- **Pastia's lines:** Bright red lines that appear in the body folds of a patient with scarlet fever after the rash develops. They are named for Constantin Pastia (1883–1926), a Romanian doctor.
- **Prodrome:** A period before the acute phase of a disease when the patient has some characteristic warning symptoms.
- **Strawberry tongue:** A swollen and intensely red tongue that is one of the classic signs of scarlet fever.

### Prognosis

The prognosis for full recovery from scarlet fever is excellent provided the child is treated with antibiotics. Untreated scarlet fever may have as much as a 3 percent chance of developing into rheumatic fever, the most troublesome complication of scarlet fever. Other possible complications of the disease include ear infections, kidney or liver damage, pneumonia, inflammation of the bones or joints, sinusitis, or meningitis (inflammation of the membranes covering the brain and spinal cord). These complications can develop within eighteen to twenty-one days after the onset of untreated scarlet fever.

Another possible complication of scarlet fever or strep throat is the PANDAS syndrome, a term that stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections. Some children develop symptoms of obsessive-compulsive disorder or Tourette syndrome after a Group A streptococcal infection, or they become moody, irritable, and anxious when separated from their parents. The connection between these symptoms and the infections is not clear, however, and many doctors think that further research is needed.

### Prevention

Scarlet fever is difficult to prevent completely because of the close contact among children in schools and day care centers. In addition, some
children (and adults) can carry Group A streptococci in their throats without getting sick themselves. When a child is diagnosed with scarlet fever, doctors recommend protecting other family members by washing the sick child's eating utensils, drinking glasses, and (if possible) toys and blankets in hot soapy water, separately from the rest of the family's dishes and laundry. The child should also be reminded to cover the nose and mouth when sneezing or coughing and to wash the hands thoroughly in warm soapy water afterward.

#### The Future

Since the introduction of antibiotics, scarlet fever is a much less serious disease than it was in the nineteenth century, when it often led to rheumatic fever and other potentially life-threatening complications.

**SEE ALSO** Ear infection; Obsessive-compulsive disorder; Rheumatic fever; Strep throat; Tourette syndrome

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# **u** Schizophrenia

# Definition

Schizophrenia is a severe, long-term, and disabling mental disorder characterized by psychosis—distorted perceptions of the real world. People diagnosed with schizophrenia suffer from delusions, hallucinations, disorganized speech and thinking, apparent lack of emotion, inability to relate to others, and significant difficulties in finishing school, holding a job, or living independently. The disorder is most likely to appear during adolescence or the early adult years.

Schizophrenia is still not completely understood by mental health professionals or medical researchers. Some doctors now think that schizophrenia is a collection of mental disorders with some common features but also symptoms that may differ from person to person, rather than a single disorder.

#### Description

Schizophrenia has existed for thousands of years but did not receive its present name until 1908. Eugen Bleuler (1857–1939), a Swiss psychiatrist, invented the word as a replacement for an older term he considered misleading. Because the word *schizophrenia* means "split mind," some people confuse it with dissociative identity disorder (formerly called multiple personality disorder). It is important to note that people with schizophrenia do not have separate internal personalities.

People with schizophrenia vary from one another in their symptom patterns and their ability to function in society. Some have periods of surprising productivity, such as John Nash (1928–), a mathematician who was hospitalized for schizophrenia in 1959 but slowly recovered during the 1980s and was awarded the Nobel Prize in economics in 1994. (A film about Nash, *A Beautiful Mind*, was released in 2001.) Other patients are never able to function well and require ongoing help from family members, psychiatrists, and social workers.

A common misunderstanding of people with schizophrenia is that they are violent. Some schizophrenics do commit bizarre or violent crimes, often under the influence of alcohol or drugs of abuse. Most people with the disorder, however, are more afraid of other people than are violent or aggressive toward them.

The role of drug and alcohol abuse in schizophrenia is complicated. On the one hand, the disorder itself is not caused by alcoholism or drug abuse. On the other, schizophrenics are more likely than others to abuse drugs and alcohol, often as a way of coping with their symptoms. One problem with drug and alcohol abuse is that it makes schizophrenia harder to treat. Drugs like phencyclidine or PCP ("angel dust"), marijuana, or cocaine can make the symptoms of schizophrenia worse; they

#### Cause

Unknown; possibly a combination of genetic, environmental, and personal factors

#### Symptoms

Delusions, hallucinations, unstable mood, lack of emotion, disorganized thinking

#### Duration

Years; usually lifelong after diagnosis

#### Schizophrenia

Normal brain image (top) and one of a patient suffering from schizophrenia. Different areas of the brain show activity, as shown by red and yellow areas. WELLCOME / PHOTO RESEARCHERS, INC.



also make patients less likely to follow their treatment plan. The drug that is most commonly abused by schizophrenics, however, is nicotine. Between 75 and 90 percent of people with schizophrenia are heavy smokers, compared with 25 to 30 percent in the general population.

# **Demographics**

Schizophrenia is thought to affect about 1 percent of the population around the world over the age of eighteen. In the United States, about 2 million people have been diagnosed with schizophrenia. As far as is known, schizophrenia affects all races and ethnic groups equally. It is considered one of the most disabling disorders in all countries, being ranked just behind spinal cord injuries and dementia in the severity of its impact on people's lives.

Risk factors for the disorder include:

- Age. Schizophrenia is largely a disorder beginning in adolescents and young adults. It is rare in children, although cases have been reported in children as young as five. It is also rare for schizophrenia to develop in adults over forty-five.
- Gender. Males are more likely to develop schizophrenia in their late teens and early twenties and to have more severe symptoms.

Females are more likely to develop symptoms in their late twenties or early thirties.

- People with a family history of schizophrenia. A child of a schizophrenic parent has a 10 percent chance of developing the disorder.
- People exposed to viruses or malnutrition prior to birth, or whose mothers had a difficult childbirth.
- People who experimented with recreational drugs during their early teens.
- People whose fathers were over forty at the time of their birth.
- People who live in large cities.

# **Causes and Symptoms**

The causes of schizophrenia are not fully understood. There is general agreement that genetic factors are involved because the disorder is known to run in families. However, no specific gene has been identified as the cause of schizophrenia. In 2007 and 2008, researchers uncovered evidence that rare genetic mutations, rather than common genes, may be responsible for the disorder—and that some of these mutations may be unique to isolated individuals rather than distributed throughout the population.

Other theories about the causes of schizophrenia include the notion that the brains of people with schizophrenia are different in structure from those of people without the disorder, or that they have abnormally high levels of a neurotransmitter (brain chemical) called dopamine. One theory that has been completely discarded in recent years is that schizophrenia is caused by bad parenting.

Doctors classify the symptoms of schizophrenia into three groups: positive, negative, and cognitive. Positive symptoms refer to behaviors that indicate a loss of contact with reality:

- Hallucinations: seeing or hearing things that are not really there. People with schizophrenia commonly have auditory (hearingrelated) hallucinations, often voices telling them to do something, including harming other people.
- Delusions. Delusions are false ideas that a person continues to hold even after they are shown to be false.
- Disorganized speech. This may include making up words that have no meaning or stringing words together in nonsensical ways.

• Movement disorders. The patient may be clumsy and uncoordinated, or may sit motionless for hours—a condition called catatonia.

Negative symptoms refer to losses or deficiencies in relating to others or in general functioning:

- · Lack of emotion in the voice or facial expressions
- Social isolation
- Neglecting personal cleanliness and grooming
- Inability to take pleasure in things that most people enjoy
- Problems in starting and organizing tasks or activities
- Refusing to speak even when spoken to

Cognitive symptoms refer to problems with thinking, memory, and the ability to pay attention. These are the symptoms that interfere most severely with the patient's education and employment in adult life.

#### Diagnosis

There is no laboratory or imaging test that can be used to diagnose schizophrenia, although in some cases the doctor may order blood or urine tests to rule out medical disorders or imaging studies of the head to rule out brain tumors or head injuries. The diagnosis is made on the basis of an interview with the doctor followed by a series of questionnaires and a detailed family history. During the interview, the doctor will listen to the patient's tone of voice and observe his or her appearance and behavior as well as the content of the answers to questions. In some cases the doctor will need to talk to members of the patient's family or close friends in order to collect information about the patient's previous ability to function; when the symptoms began; and whether there is a family history of mental illness.

To be diagnosed with schizophrenia, the patient must meet the following criteria:

- The presence of at least two of the following: delusions, hallucinations, disorganized speech, catatonic behavior, or any negative symptoms.
- Significant inability to function at home, in school or at work, or in carrying out daily tasks.
- The symptoms have lasted for at least six months.
- Other mental health disorders have been ruled out.

# Treatment

The treatment of schizophrenia is a lifelong process; there is no cure for the disorder. Some patients occasionally require hospitalization if they appear to be dangerous to themselves or others. Most, however, can be treated as outpatients. One of the chief difficulties with therapy for schizophrenia, however, is that most patients do not think that they are ill or that they need treatment. Doctors describe this characteristic as a lack of insight into the illness.

The mainstay of treatment for schizophrenia is medications, known as anti-psychotics. These are divided into two categories: older drugs that have been used since the 1950s and newer drugs introduced in 1989. The older drugs include medications like Haldol and Thorazine, which often had severe side effects and were also not effective in treating such symptoms as the lack of motivation or feeling. The newer drugs, which are sometimes called atypical anti-psychotics, include medications like Risperdal and Zyprexa. They also have side effects. Whichever medication is prescribed, however, it is critical for patients with schizophrenia to take their medications correctly and regularly. Failure to take the medications is the most common reason for relapses.

In addition to medications, patients with schizophrenia need psychotherapy and other forms of help:

- Individual therapy. This form of treatment is recommended to help schizophrenics understand their disorder, realize the importance of taking their medications, and learn to handle everyday life problems.
- Family therapy. Some doctors recommend family therapy as a way to help family members cope more effectively with the stresses produced by caring for someone with schizophrenia.
- Rehabilitation. Rehabilitation includes training in job-related as well as social skills so that patients are better equipped to live independently.
- Treatment for co-occurring substance abuse. Between 30 and 70 percent of patients with schizophrenia are also diagnosed with alcoholism or substance abuse disorders. Patients usually do better in programs that combine drug treatment with treatment for schizophrenia than in programs that treat the disorders separately.

## WORDS TO KNOW

**Anti-psychotics:** A group of drugs used to treat schizophrenia. The older anti-psychotic drugs are also called neuroleptics.

Auditory: Pertaining to the sense of hearing.

**Catatonia:** A condition in which a person sits motionless for long periods of time and does not respond to others.

**Delusion:** In medicine, a false belief that a person holds to despite evidence or proof that it is false.

**Hallucination:** Perceiving something that is not really there. Hallucinations can affect any of the five senses.

**Psychosis:** Severe mental illness marked by hallucinations and loss of contact with the real world.

**Relapse:** Recurrence of an illness after a period of improvement.

• Self-help groups. Patients with schizophrenia benefit from the opportunities to learn and practice social skills in these groups as well as find support for the problems they face.

#### Prognosis

The newer anti-psychotic medications have helped many patients with schizophrenia who did not respond to the older drugs. However, there are still some who do not respond to medications at all or choose to stop taking them after a year or so because of the side effects. Despite ongoing research, schizophrenia remains a difficult disease to treat, and the prognosis for recovery is still poor. A few patients do recover completely, but most have periodic relapses and need group homes or long-term structured programs in order to function in the community. In addition to problems with drug abuse and dependence, patients with schizophrenia also have a very high rate of suicide—10 percent.

Patients who have a family history of schizophrenia, whose symptoms began in their teens, and who have negative symptoms have the poorest prognosis. Patients who were older when their symptoms started, got treatment quickly, and had a high level of functioning before they were diagnosed have the most hopeful prognosis for recovery.

#### Prevention

There is not enough known about the causes of schizophrenia for any preventive strategies to be effective.

# The Future

Most researchers in the field of mental health do not expect schizophrenia to become either more or less common than it is at present. They are focusing on behavioral treatments for schizophrenia as well as newer medications with fewer side effects. One new drug currently in clinical trials is administered as a nasal spray rather than an injection; another is a drug that needs to be given only twice a month rather than every day, which would make it easier for patients to stick with medication therapy. Another critical area of research is the genetic factors involved in the disorder.

SEE ALSO Alcoholism; Bipolar disorder; Marijuana use; Smoking

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# **m** Scoliosis

# Definition

Scoliosis is an abnormal curvature of the spine from side to side. It may also include an abnormal rotation of the spine. There are two basic types of scoliosis: idiopathic, which is the more common and accounts for 80–85 percent of cases; and scoliosis that is caused by a disease, injury, or temporary condition. Idiopathic means that the cause of the disorder is unknown.

The 15–20 percent of cases of scoliosis that do have causes are divided into two categories, structural and nonstructural (also called functional). A child with nonstructural or functional scoliosis has a spine that is structurally normal but looks curved at the moment because of a difference in leg length or muscle spasms. It can be treated by therapy for the underlying cause. Structural scoliosis is caused by certain neuromuscular diseases, including Marfan syndrome, muscular dystrophy, and polio; or by infectious diseases or tumors on the spinal column. In adults it may be caused by osteoporosis or compression fractures.

#### Also Known As Curvature of the spine

#### Cause

Birth defects, other diseases, unknown causes

#### Symptoms

Uneven gait, uneven shoulders or hips, protruding shoulder blades, back pain

#### Duration

Lifelong unless corrected

#### Description

Scoliosis is an abnormal curvature of the spine from side to side and usually rotated inward or outward as well. It is best understood as a complex three-dimensional distortion of the spine's normal pattern. The normal human spine does have an S-shaped curve when viewed from the side; that is, the twenty-five vertebrae in the human spinal column are not arranged in a straight line from the neck to the tailbone but curve inward slightly in the neck region, outward in the chest area, and inward again in the lower back. When viewed from the center of the patient's back, however, the vertebrae should lie in a straight line. Any curvature of 10 degrees or more to the left or right is considered scoliosis.

#### Scoliosis

*Illustration of a spine abnormally curved in scoliosis.* © PHOTOTAKE INC. / ALAMY.



The severity of scoliosis varies; some children do not require treatment at all and others do well simply having their growth and any changes in the spine monitored by their doctor. Still others require treatment, most often with either specially designed braces or surgery. Adults with scoliosis are more likely than children to have deformities severe enough to cause noticeable pain and difficulties with breathing.

#### **Demographics**

Scoliosis can occur at any age, but most studies of it focus on children and adolescents, which are the age groups most commonly affected. According to the National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS), between three and five out of every 1,000 children in the United States have scoliosis severe enough to require treatment. Scoliosis is most likely to affect children over the age of ten. Babies who are born with scoliosis are said to have congenital scoliosis.

In young children, scoliosis affects boys and girls equally; in adolescence, however, girls are twice as likely as boys to develop scoliosis. The curvature of the spine generally worsens during growth spurts.

Curves that face the right side of the child's body are more common than those that face left, and C-shaped curves are more common than S-shaped curves. In adults, scoliosis may result from a condition that actually began in childhood and was not diagnosed or treated while the person was still growing. What may have started out as a slight or moderate curve in the spine may have become worse in the absence of treatment. In other cases, adult scoliosis results from osteoporosis or other disorders that affect an older person's bones and muscles.

#### **Causes and Symptoms**

Scoliosis may have a number of different causes.

- Abnormal development of the spine prior to birth. Most cases of congenital scoliosis are caused by either abnormally shaped vertebrae or fusion of the baby's ribs.
- Genetic factors. Although only one specific gene has been linked to scoliosis, there are likely to be others involved. The disorder is known to run in some families. A person with a family member who has scoliosis has a 20 percent greater chance of developing scoliosis themselves.
- Injury. Some cases of scoliosis are the result of injuries to a baby's spine during a difficult childbirth.
- Diseases that affect connective tissue, the muscles, or the skeleton. These include polio, Marfan syndrome, muscular dystrophy, or cerebral palsy.
- Tumors on the spinal column.
- Infectious diseases.
- Osteoporosis and other diseases that may weaken bones in older adults.

The symptoms of scoliosis vary in severity from child to child; some common symptoms include:

- Protruding shoulder blades or a hump near the rib cage (caused by rotation of the spine inward)
- Uneven development of the back muscles
- Uneven hip and shoulder levels
- Uneven development of the breasts in adolescent girls
- Fatigue or pain in the spine after long periods of sitting or standing
- Unequal distance between the arms and the body
- Head may be tilted off-center
- Tendency to walk with a rolling gait

In adults, scoliosis is more likely to be associated with the development of a hump in the upper back, breathing disorders, and pain.

#### Diagnosis

Diagnosis of scoliosis begins with a family history as well as a history of the child's medical problems, birth defects (if any), and injuries that may have affected the spine. The doctor will also examine the child's skin for café-au-lait spots; these are brownish-white spots that indicate the scoliosis may be the result of a birth defect. If the patient is an adult, the doctor will look for evidence that the scoliosis is the result of a childhood problem that was never treated, or that it is due to an adult-onset disorder like osteoporosis.

The most common diagnostic test for scoliosis in adults as well as children is the Adam Forward Bending Test. The patient faces forward with feet straight ahead and palms inward against the sides of the body. With the knees locked, the patient slowly bends over at the waist and tries to touch the toes. The doctor then evaluates the spine for the appearance of straightness. An x ray may be taken to allow the doctor to measure the exact degree of abnormal curvature. X-ray photographs can also be used to measure the improvement in the spine during treatment.

Adult patients may be given a neurological examination as well as an x ray if their scoliosis is causing difficulties in bowel or bladder habits, general weakness, or unusual sensations in the arms or legs.

#### Treatment

Many children are diagnosed with mild scoliosis as part of a school screening program and may not need special treatment. If the child's doctor thinks that he or she should be treated, the child will usually be referred to an orthopedic specialist.

Treatment of scoliosis is based on the degree of the curvature of the patient's spine; his or her age; whether the patient is likely to continue growing; and the type or cause of the scoliosis:

• Observation. If the child is still growing, has idiopathic scoliosis, and has a curvature of 25 degrees or less, the doctor will usually check the child's growth during an office visit every four to six months.

- Bracing. If the child has at least two years of growth left; is a girl who has not yet had her first period; has idiopathic scoliosis with a curve greater than 25–30 degrees; or has a curve that is getting noticeably worse, the doctor will recommend braces. A child who must wear a brace can, however, participate in a full range of school and social activities. If properly fitted, braces can prevent the need for surgery in 90 percent of cases.
- Surgery. Surgery may be needed when the child has stopped growing; has a curvature that is greater than 45 degrees; or has a curvature that is getting worse rapidly. Surgical treatment of sco-liosis involves fusing several vertebrae together to correct the curvature and may require inserting metal rods next to the spine to reinforce the fusion.

Adult patients with scoliosis are usually treated with surgery either for pain control or to relieve pressure on the heart and lungs from the deformed spine.

Other forms of treatment for scoliosis that are not considered effective include dietary changes, vitamin supplements, exercise programs, chiropractic manipulation of the spine, and electrical stimulation.

Although physical exercise should not be used as a treatment to correct the curvature of the spine, it is an important part of overall health care for children with scoliosis. Regular exercise helps to keep the body fit and healthy, improves overall well-being, and reduces the risk of osteoporosis in girls and women.

#### Prognosis

The prognosis of scoliosis depends on the risk that the curvature of the patient's spine will progress (get worse). In general, the prognosis is better for smaller curves than for larger ones, and better for patients whose bones are mature than for those whose spines are still growing. In general, mild cases of scoliosis treated with bracing alone do very well. These patients do not usually have long-term health problems except for a slightly increased risk of pain in the lower back when they get older. People with surgically corrected idiopathic scoliosis also do very well and can lead active, healthy lives.

The prognosis of patients with scoliosis related to neuromuscular diseases like muscular dystrophy or cerebral palsy is determined by the outcome of their disease rather than by treatment for the scoliosis by itself.

# WORDS TO KNOW

**Café-au-lait spots:** Brownish-white birthmarks that appear as part of a nervous system disorder that can cause scoliosis in some children. Café au lait is the French expression for "coffee with milk" and describes the color of the spots.

**Chiropractic:** An approach to treatment of disease based on manipulation of the spinal column.

Congenital: Present at birth.

Gait: A person's habitual manner of walking.

**Idiopathic:** The medical term for a disorder whose cause is unknown.

**Orthopedics (also spelled orthopaedics):** The branch of medicine that deals with the prevention, diagnosis, and treatment of disorders of the skeleton and muscles.

# Prevention

Scoliosis cannot be prevented in the great majority of cases because its causes are still not fully understood.

# **The Future**

Doctors expect that the number of cases of adult-onset scoliosis will increase in the years ahead because of the growing number of people over 65 in the general population. Earlier diagnosis and treatment of scoliosis in children, however, is lowering the need for surgery as a treatment for the disorder.

After the discovery in 2007 of the first gene linked to scoliosis, it is likely that researchers will identify other genes related to the condition. These discoveries in turn may shed light on the causes of idiopathic scoliosis and possibly lead to the development of new treatments.

**SEE ALSO** Cerebral palsy; Marfan syndrome; Muscular dystrophy; Osteoporosis; Polio

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# m

# Seasonal Affective Disorder

## Definition

Seasonal affective disorder, or SAD, is a form of depression related to the changes in the seasons. Most people with SAD feel tired, depressed, and uninterested in life during the fall and winter and feel happier and more energetic in spring and summer. There are, however, some people who follow the opposite pattern, feeling depressed in spring and summer and livelier in fall and winter. This summer pattern is sometimes called summer SAD, reverse seasonal affective disorder, or RSAD.

#### Description

Seasonal affective disorder has troubled humans, particularly those living at some distance from the equator, for centuries. It used to be part of common folk wisdom that the changing seasons affect the way people feel. With the coming of electric lighting and indoor climate control, however, modern people are less in touch with the influence of seasonal changes in temperature and light intensity on human moods. Dr. Thomas Wehr, an expert on SAD, commented in an interview in 2002 that "We've kind of de-seasonalized ourselves as much as possible. You know, we turn the lights on after dark, we turn the heat on in winter, we turn the air conditioning on in summer, and you could almost not notice [the weather]."

#### Also Known As

SAD, winter blues, winter depression

#### Cause

Unknown; may be partly genetic

#### Symptoms

Depression related to changes in seasons

#### Duration

Two to four months out of every year

#### Seasonal Affective Disorder



Pilots in the Hensinki airport using bright lamps to combat seasonal affective disorder. © PICTURE CONTACT / ALAMY.

The symptoms of winter SAD vary in intensity in the general North American population. Some people are only mildly affected, feeling some loss of energy during the winter, a tendency to oversleep and gain weight, an increased craving for starchy foods, and lowered interest in social activities, but others feel severely depressed to the point of requiring hospitalization. The symptoms of RSAD are somewhat different, with people feeling agitated or jumpy, finding themselves having difficulty sleeping, and in some cases, thinking about suicide.

#### **Demographics**

Researchers at the National Institute of Mental Health (NIMH) estimate that as many as 5 percent, or 15 million Americans suffer severely from SAD, with another 33 million feeling some moodiness or loss of creativity or productivity during the winter. There appears to be some connection between geographical location and the risk of SAD, with the rates of winter SAD increasing from south to north in the United States. About 1.5 percent of people in Florida have winter SAD, compared to 9 percent in Minnesota and Maine. Areas with overcast skies in the winter also have higher rates of SAD than those with relatively clear winter weather.

Women are about twice as likely as men to suffer from SAD. The disorder is most likely to appear in the early adult years, but some patients report symptoms of SAD going back to childhood.

# A Doctor's SAD Story

Dr. Norman Rosenthal, the psychiatrist who first identified seasonal affective disorder (SAD), grew up in South Africa, where the length of daylight at different times of the year does not vary as much as it does in most parts of the United States. In the summer of 1976 Rosenthal moved to New York City and was surprised by the difference between the energy he enjoyed during the warm months and a tiredness that began almost as soon as daylight saving time ended. He moved in 1977 to the National Institute of Mental Health (NIMH) in Maryland and met several patients who seemed to suffer from depression related to the seasons.

Rosenthal and several colleagues experimented with having these patients sit in front of boxes containing light bulbs covered by a plastic screen for several hours a day and were surprised by how quickly the patients responded to light therapy. At first the doctors thought this seasonal type of depression might be very rare. They decided to contact the health journalist at the Washington Post to see whether there might be other people in the Washington area with the condition. They received thousands of phone calls from people all over the United States who were eager to participate in research that might help them. By 1981 Rosenthal recognized that his first winter in the United States was an indication of his own mild form of SAD.

About 85 percent of patients with SAD have the winter blues, with the remaining 15 percent having summer SAD. Summer SAD is more common in hotter climates, such as India, parts of China, or the American South.

#### **Causes and Symptoms**

The causes of SAD are not completely understood, but there are several theories that have been proposed to explain the disorder:

- Genetic factors. About two-thirds of people diagnosed with SAD have at least one relative with depression or another mood disorder.
- Abnormal levels of melatonin. Melatonin is a hormone produced by the pineal gland in the brain that regulates the daily circadian rhythm (sleep/wake cycle) in humans. Humans generally produce more melatonin during the long nights of winter, and some researchers think that overproduction of melatonin in some people may be responsible for the depressed mood of SAD.
- Disruption of the circadian rhythm caused by the decreasing levels of sunlight in fall and winter. There are some researchers who think that SAD is related to jet lag, another form of depression that is associated with disruption of a person's usual sleep/wake pattern caused by traveling across too many time zones too rapidly.
- Lack of serotonin. Serotonin is a neurotransmitter (chemical produced by the

brain) that is known to affect mood. The lower levels of sunlight in the winter are thought to cause a drop in serotonin production, which in turn leads to depression. The causes of summer SAD are less clear-cut. Some doctors think that it may be more related to summer heat than to seasonal changes in light levels.

The symptoms of winter SAD typically include:

- Oversleeping, having difficulty getting up in the morning.
- Craving for heavy, starchy foods and sweets. Some people with winter SAD gain so much weight every winter and lose it in the summer that they have wardrobes in two different sizes for the different seasons.
- Lack of interest in work or social activities.
- Feelings of depression and hopelessness.
- Difficulty concentrating or paying attention.

The symptoms of summer SAD include:

- Anxiety
- Loss of appetite and weight loss
- Insomnia
- Agitation
- Irritable mood

#### Diagnosis

The diagnosis of SAD is based on the doctor's discussion of the patient's symptoms with him or her and completion of a set of questionnaires about SAD. The patient will usually be asked about seasonal changes in mood and behavior, his or her lifestyle and social situation, and sleeping and eating patterns. In most cases the doctor will also give the patient a physical examination to rule out changes in mood that could be caused by thyroid disease or other disorders.

#### Treatment

Treatment for winter SAD usually includes light box therapy, in which a person sits in front of a box containing bright white lights covered by a plastic filter to prevent glare. Treatments usually last for thirty to sixty minutes in the morning (or whatever time of day works best for the patient). The patient is instructed to sit near the box with eyes open but without staring at the box. People who find light boxes inconvenient are sometimes helped just by taking walks outside in the winter during daylight hours. One advantage of light therapy is that it is easy to use and has few side effects.

Other treatments for winter SAD include antidepressant medications; timed doses of melatonin in the early evening to shift the body's circadian rhythm; and psychotherapy to help people cope with the mood changes of winter SAD.

Some people with summer SAD are helped simply by staying in airconditioned environments as much of the time as they can, but others need antidepressant medications as well.

#### Prognosis

The prognosis of SAD depends on the severity of the patients' symptoms and whether they have another mental disorder in addition to SAD. People with only mild symptoms often do well on their own by using more bright lights in their homes in the winter and spending more time outside.

Between 50 and 80 percent of patients with severe SAD benefit from light box therapy, and about 67 percent of patients feel better when treated with antidepressants. Timed doses of melatonin appear to be less effective with most people.

Patients who suffer from major depression as well as SAD should be treated for both disorders. About 20 percent of patients with severe SAD are eventually diagnosed with bipolar disorder.

# Prevention

There is no known way to prevent SAD, but learning about the condition and beginning treatment each year before the seasonal symptoms get underway can keep them from getting worse as the season progresses. Sticking to a treatment plan and getting regular physical exercise are also ways to minimize the effects of SAD on one's life.

# The Future

Researchers are hopeful that better understanding of the causes of both winter and summer SAD will lead to more effective treatments, as none of the present therapies work for all patients with the disorder.

SEE ALSO Bipolar disorder; Depression

## WORDS TO KNOW

**Circadian rhythm:** The medical name for the daily sleep/wake cycle in humans.

- Jet lag: A sleep disorder or disturbance in the sleep/wake cycle related to rapid travel across time zones.
- **Melatonin:** A hormone produced in the pineal gland in the brain that regulates the sleep/wake cycle.
- **Serotonin:** A chemical produced in the brain that regulates mood.

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# **m** Seizure Disorder

# Definition

Seizure disorder, also called epilepsy, is a disorder of the brain in which there is an abnormal brief surge of the electrical impulses that travel between brain cells. Instead of the normal pattern of signal transmission, the nerve cells misfire as many as 500 times a second, much faster than usual. The affected person's symptoms may range from a few seconds of blank staring to several minutes of convulsions, loss of consciousness, or muscle spasms.

One in every ten adults around the world will have a seizure at some point in his or her life. To be diagnosed with seizure disorder, however, a person must have two or more seizures.

# Description

Seizure disorder has been known to doctors for thousands of years. The loss of control, strange movements, and unpredictable behaviors of people having seizures caused some ancient writers to wonder whether they were possessed by demons. Eventually, however, Hippocrates in the fifth century BCE attributed seizures to a problem within the brain. His insight proved to be correct. It is important to keep in mind that seizure disorder is not caused by a mental disorder or by mental retardation. Although some mentally retarded people do have seizures, having seizures does not mean that the affected person is or will become mentally retarded.

Doctors divide seizures into four large categories:

- Generalized seizures (sometimes called grand mal seizures) affect both sides of the brain and include: absence seizures, in which the person stares into space briefly; atonic seizures, in which the person goes limp or falls down; and tonic-clonic seizures, in which the person may lose consciousness, stiffen the body, or make jerking movements of the arms and legs.
- Focal seizures occur in only one part of the brain and affect about 60 percent of people diagnosed with seizure disorder. In focal seizures, the person may have hallucinations or other sensory

#### Also Known As Epilepsy

#### Cause

Brain abnormalities, genetic factors, stroke, head injury, poisoning, infectious diseases

#### Symptoms

Blank staring, loss of consciousness, convulsions, jerking of arms and legs

#### Duration

A few seconds to a few minutes



Four-year-old girl undergoing an electroencephalogram (EEG) to diagnose a seizure disorder. AJPHOTO / PHOTO RESEARCHERS, INC.

disturbances or experience sudden but intense emotions. In some cases, a person having a focal seizure may perform repetitious movements like eye blinking, mouth twitching, or walking in a circle.

- Nonepileptic seizures may look like focal or generalized seizures, but are not caused by electrical disruptions in the brain. They may be caused by a high fever, strong drugs, general anesthesia, or certain complications of pregnancy.
- Status epilepticus is the medical term for an ongoing seizure that lasts longer than five minutes or seizures that follow each other without the individual waking up. It is potentially life-threatening and needs emergency treatment.

It is possible for a person with seizure disorder to have more than one type of seizure.

Some people with seizure disorder find that their seizures are triggered by certain conditions or activities; others do not have recognizable triggers. The most common trigger for a seizure is failure to take prescribed antiseizure medication. Other triggers include heavy drinking, lack of sleep, emotional stress, or (in women) hormonal changes associated with the menstrual cycle. Seizure triggers do not cause seizures in the strict sense, they simply set them off.

# First Aid for a Seizure

A generalized seizure can be frightening for onlookers to see; however, most are not medical emergencies. The Epilepsy Foundation recommends the following steps to help a person having a seizure:

- Stay calm and calm down other people nearby.
- Do not try to hold the person or stop his or her movements.
- Time the seizure with a nearby clock or watch.
- Remove any hard or sharp objects near the person.
- Loosen his or her tie or collar.
- Put a folded towel or sweater under the person's head and turn the head to one side to keep the airway clear.
- Do not try to force the person's mouth open.
- Do not try to give the person artificial respiration.
- Stay with the person until he or she regains normal consciousness.
- Be friendly and reassuring.
- Offer to call a taxi or family member to take him or her home.

If the seizure lasts longer than five minutes, the person stops breathing, the seizure happened in the water, or the person appears to be injured in any way, call 911 at once.

# **Demographics**

Doctors estimate that about one person in every 100 around the world, or about 50 million people in all, has seizure disorder. In the United States, about 200,000 people are diagnosed with seizure disorder each year, 45,000 of them children below the age of fifteen. There is no apparent cause of the seizures in over half of newly diagnosed cases.

About half of newly diagnosed patients have generalized seizures. Generalized seizures are more common in children under the age of ten than in adults.

Risk factors for seizure disorder include:

- Age: Children younger than two years and adults over sixty-five are more likely to develop the condition.
- Sex: Males are slightly more likely to develop seizure disorder than females.
- Race: African Americans are more likely to develop seizure disorder than members of other racial groups.

The rate of seizure disorder is higher in those with other disorders that affect the nervous system:

- 10 percent of patients with Alzheimer disease
- 22 percent of patients with stroke
- 10 percent of children with cerebral palsy
- 10 percent of children with mental retardation
- 8.7 percent of children whose mothers have seizure disorder
- 2.4 percent of children whose fathers have seizure disorder

# **Causes and Symptoms**

Seizure disorder can have a number of different possible causes. A few rare types of epilepsy have been traced to specific genes; a few other types are

known to run in families, though they have not been linked to specific genes. In some cases, seizures are a result of head injuries, cerebral palsy, autism, Alzheimer disease, alcohol abuse, brain tumors, AIDS, and other infectious diseases that affect the brain. In about 50 percent of cases, however, doctors cannot identify a specific cause of the patient's seizure disorder.

Symptoms of seizure disorder vary depending on the type of seizure:

- Generalized absence seizures (sometimes called petit mal seizures): The patient stares off into space and appears to be "out of it" or inattentive.
- Generalized tonic-clonic seizures: These are the dramatic generalized seizures that many people picture in their mind when they think of a seizure. The person may fall on the ground, lose consciousness, thrash about, and lose bowel or bladder control. In these types of seizures, after the jerking ends, the patient will be asleep or drowsy, which is called the post-ictal state.
- Generalized atonic seizures: The person goes limp and may slump (if sitting) or fall down (if standing).
- Generalized myoclonic seizures: The person makes sudden jerking or twitching movements of the arms and legs.
- Simple focal seizures: In a simple focal seizure, there is no change in the patient's level of consciousness. The patient may experience a sudden strong emotion or notice changes in the way things look, sound, taste, or feel.
- Complex focal seizures: A complex focal seizure is one in which the patient loses consciousness for a few moments, although he or she may continue to make purposeless repetitive movements like lip smacking, swallowing, or picking at clothing.

Many people do not understand enough about seizure disorder and its wide range of possible symptoms to respond appropriately to a person having a seizure. It is not unusual for people having nonconvulsive seizures to be treated as if they are mentally ill. Thus widespread lack of understanding is one of the biggest social problems for people with seizure disorder.

#### Diagnosis

Seizure disorder can be difficult to diagnose, because seizures can be caused by meningitis, encephalitis, or a stroke, which all need emergency attention. The person having the seizure may not remember what happened when he or she returns to normal consciousness, and a family member or bystander may need to describe to a doctor the patient's symptoms and whether there have been previous seizures. After any necessary emergency medical treatment is given, the next step in diagnosis is a neurological examination to test the patient's reflexes, sight, hearing, muscle tone, gait, posture, balance, coordination, and ability to talk normally and answer simple questions.

The specific laboratory tests and imaging studies that may be ordered depend on the specific symptoms associated with the seizure:

- Blood tests to look for evidence of infection, diabetes, or anemia.
- Electroencephalogram (EEG) to record the patterns of electrical activity in the patient's brain. In some types of seizure disorder, the patient's brain waves will be abnormal even when he or she is not having a seizure.
- Computed tomography (CT) scan, magnetic resonance imaging (MRI), or positron emission tomography (PET) scans of the head to identify brain tumors, evidence of a stroke, or other structural abnormalities of the brain. PET can also be used to identify the parts of the brain responsible for focal seizures.

The test results are used to determine which type of seizure disorder the patient has and if possible, the likely cause of the seizures, which helps to guide treatment decisions.

#### Treatment

Antiseizure medications are the first line of treatment for seizure disorder. Most patients need only one medication, but some may need a combination of two or more. It may take several trials of different drugs to determine which one works best and what dosage is most effective.

Some patients whose seizures are caused by a small portion of the brain may benefit from surgery, but only if the affected part of the brain does not control sight, hearing, or other vital functions.

Some patients are also helped by a vagus nerve stimulator, a device implanted beneath the collarbone near the vagus nerve in the neck. It is not clear why stimulation of this particular nerve helps to control seizures, but it is reported to lower the number of seizures by 20–40 percent in most patients.

## WORDS TO KNOW

**Intractable:** Referring to a disease or disorder that cannot be easily treated or cured.

**Status epilepticus:** An ongoing seizure that lasts longer than five minutes; it is a medical emergency.

#### Prognosis

The prognosis of seizure disorder varies according to the patient's age at the time of the first seizure as well as the type of seizure. About 80 percent of people with seizure disorder can be successfully treated with medications; the remaining 20 percent are said to have intractable (difficult-to-treat) epilepsy. About 75 percent of people who are seizure-free on medication for two to five years can be successfully withdrawn from medication. Seizure disorder does carry with it, however, an increased risk of sudden unexplained death or of status epilepticus, from which about 42,000 people die annually in the United States.

Some people with seizure disorder have difficulty finishing school and with employment. One reason is social misunderstanding; another is the effect of antiseizure drugs on a person's ability to concentrate. Employment difficulties are usually related to the restrictions that most states place on driving. Most states require a person with a history of seizure disorder to show that he or she has been seizure-free for a specified period of time before he or she can apply for a driver's license.

#### Prevention

Seizure disorder is difficult to prevent, given that it has so many different forms and the fact that doctors cannot identify a cause in about half of all cases. The best way that patients diagnosed with the disorder can prevent seizures is to take their prescribed medications as directed.

#### The Future

Present research on seizure disorder is focused on improving brain imaging techniques so that doctors will be better able to identify patients who can be helped by surgery as well as medications for seizure disorder. Other researchers are looking for additional genes that may be related to seizure disorder or studying the effectiveness of special diets in treating seizures. **SEE ALSO** AIDS; Alcoholism; Alzheimer disease; Brain tumors; Cerebral palsy; Concussion; Encephalitis; Meningitis; Stroke

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# Severe Acute Respiratory Syndrome

# Definition

Severe acute respiratory syndrome, or SARS, is a potentially lifethreatening infection of the upper respiratory tract caused by a new type of virus that had not been seen before the fall of 2002. This new virus, which has been named the SARS-associated coronavirus (SARS-CoV), belongs to a family of viruses called coronaviruses because they have a crown-shaped appearance under a microscope. Before the SARS outbreak of 2002–2003, coronaviruses were thought of largely as a cause of the common cold. The SARS virus was identified as the cause of the disease by a researcher at the University of Hong Kong in March 2003.

SARS is defined by both WHO and the CDC as an emerging disease. This classification means that SARS is considered to be a disease that has become more widespread around the world in the last twenty years and may become more common in the future.

# Description

The first descriptions of SARS indicated that most patients thought at first that they had influenza. The early symptoms of the disease often include such flu-like symptoms as chills, sore muscles, fever, and a generally unwell feeling. In the second phase of the illness, the person develops a dry cough that can develop into pneumonia and difficulty breathing. At that point the person may need mechanical ventilation on a respirator.

# **Demographics**

SARS does not appear to strike any specific age group more frequently than any other. However, the elderly are at greatest risk of death from the disease. As far as is known, men and women are equally affected, as are people of all races and ethnic groups.

The country most severely affected by the 2002–2003 SARS epidemic was mainland China, with 5,327 confirmed cases and 349 deaths. In Hong Kong there were 1,755 cases and 299 deaths. The outbreak around Toronto, Canada, which involved 251 cases and forty-three deaths received considerable media attention and prompted widespread fear of the disease across North America. As of July 1, 2007, the CDC reported that there have been eight confirmed cases of SARS in the United States; all the patients had been traveling in countries where SARS had been reported. All eight patients survived.

One of the strange aspects of the SARS epidemic is that the disease has virtually disappeared since late 2004. The CDC reported on May 3, 2005, that "there is no known SARS transmission anywhere in the world" as of that date. The last known cases of SARS were reported in a group of Chinese

Also Known As SARS

**Cause** Coronavirus

#### Symptoms

Severe difficulty in breathing; influenza-like symptoms (muscle pains, cough, fever)

**Duration** Two to three weeks



Illustration of a SARS virus. The wavy line in the middle of the cell is its genetic material. JIM DOWDALLS / PHOTO RESEARCHERS, INC. laboratory workers in April 2004. The WHO SARS Web site has not been updated since October 2004.

## **Causes and Symptoms**

SARS is caused by a coronavirus belonging to the same family of viruses responsible for some common colds. The first known case of SARS occurred in a farmer in Guangdong Province, China, in November 2002. The farmer died shortly after being taken to the hospital. After the SARS virus was found in civets (small animals resembling otters or skunks) and other animals sold for food in the markets around Guangdong in May 2003, some researchers concluded that SARS is a zoonosis, or disease that people can get from animals. Almost all reported cases, however, involve person-to-person transmission through droplets released into the air when an infected

person coughs or sneezes, or by direct contact with a drinking glass or other object touched by an infected person. The SARS virus also appears to remain in the air as small particles for some time after an infected person has sneezed. This method of transmission means that someone can get SARS even when he or she is not face to face with someone who has the disease.

SARS has an incubation period that can vary from two to ten days. The patient then runs a fever of around 100.4°F (38°C) or higher and experiences muscle aches, headache, and other influenza-like symptoms. About 20 percent of patients develop nausea and vomiting; some develop diarrhea as well.

The second phase of SARS begins about three days after the person first feels sick. The person develops a dry cough, has difficulty breathing, may wheeze, and begins to suffer from lack of oxygen in the blood. Even if the patient is helped by being placed on a respirator, he or she may die.

# Diagnosis

The diagnosis of SARS is usually based on a combination of the patient's history—particularly travel within the previous ten days to a country with reported cases of SARS—and the fever. Chest x rays do not always show anything unusual during the first week of infection, and the doctor may not hear anything abnormal when he or she listens to the patient's breathing.

# A Medical Pioneer's Courage

Dr. Carlo Urbani (1956-2003) was a specialist in infectious diseases who worked for the World Health Organization (WHO). When the Vietnam French Hospital in Hanoi contacted WHO on February 28, 2003, about a patient who was seriously ill with an unidentifiable influenza-like virus, Urbani went immediately to Hanoi. Looking back, doctors now know that the hospital was trying to cope with an outbreak of SARS. Of the first sixty patients who had the new disease, more than half were doctors and nurses. In addition to carrying out the measures that were needed to control infection inside the hospital and sending samples for laboratory testing, Urbani recognized the seriousness of the situation and notified the Vietnamese government of the need for urgent public health measures. The government of Vietnam took the unusual step of calling for outside assistance rather than giving priority to its public image, as had happened in China.

Urbani flew to Bangkok, Thailand, on March 11 to meet an American doctor from the Centers for Disease Control and Prevention (CDC). During the flight he began to feel sick. Urbani was placed in an isolation room in a Bangkok hospital but died of SARS on March 29. He was only forty-six years old.



Carlo Urbani. AP IMAGES

The Italian chapter of Doctors Without Borders, which had elected Urbani to be its president in 1999, credits him with saving the lives of thousands of people around the world by his timely and courageous warning to WHO and other public health organizations about the new disease.

When SARS first emerged, doctors had to make the diagnosis on the basis of the fever and the patient's history of travel or of close contact with an infected person. After the genome of the SARS virus was successfully sequenced (analyzed) by Canadian scientists in April 2003, it was possible to develop laboratory tests to detect antibodies to the virus and also take the first steps toward developing a vaccine. As of 2008, there were three types of diagnostic tests that can be used to detect SARS:

• A DNA rapid polymerase chain reaction test that uses a sample of blood or nasal secretions to check for the DNA of the SARS virus.

- A blood test that checks the patient's blood for the presence of antibodies to the SARS virus.
- A viral culture. In this type of test, a small amount of tissue fluid from the patient or small piece of tissue is incubated for a period of time in a special culture medium and then analyzed for measurable amounts of the SARS virus.

# Treatment

Treatment for SARS is largely supportive, as antibiotics are not effective in treating viruses. Although steroid medications have been used in Hong Kong and a few other locations to treat SARS patients, it is not certain whether these medications are effective. Patients are primarily treated in critical care units where they can be placed on a respirator if needed, and they are kept in strict isolation from other patients.

Patients who may have SARS should tell their doctors who their contacts have been within the past ten to fourteen days, so that these people can be evaluated for early signs of the disease.

# Prognosis

The prognosis for recovery from SARS depends largely on the patient's age and general health. In the pandemic of 2002–2003, the highest mortality rates (over 50 percent) were among people over the age of sixty-five, whereas the overall fatality rate for the outbreak was 9.6 percent.

# Prevention

The most important aspect of preventing SARS is keeping patients known to have the disease in isolation from others. Other preventive measures recommended by WHO and the CDC include the following:

- Before traveling abroad, check with the CDC for any recent reports of SARS in other countries. When traveling, carry a basic first-aid kit that includes an alcohol-based hand cleanser.
- At home as well as abroad, wash the hands frequently with hot soapy water.
- If caring for a person with SARS, use disposable gloves and throw them away after use. Never try to reuse the gloves. Wear a surgical mask when in the same room as the patient.

# WORDS TO KNOW

**Bioterrorism:** The use of disease agents to frighten or attack civilians.

- **Emerging infectious disease (EID):** A disease that has become more widespread around the world in the last twenty years and is expected to become more common in the future.
- **Pandemic:** A disease epidemic that spreads over a wide geographical area and affects a large proportion of the population.

**Zoonosis:** A disease that animals can transmit to humans.

- Wash the patient's dishes, towels, bedding, silverware, and clothing in hot water apart from the rest of the family's dishes or laundry.
- Use chlorine bleach or another household disinfectant to clean any surfaces that may have been contaminated with the patient's sweat, saliva, mucus, vomit, stool, or urine, as the SARS virus can survive for several days in human body fluids.
- Follow these safeguards for at least ten days after the last of the patient's symptoms have cleared up.

#### **The Future**

Work continues on the development of a vaccine against SARS as well as the development of possible treatments. The National Institutes of Health (NIH) has been conducting clinical trials of a SARS vaccine since 2004. Some researchers are investigating the possibility that the antiviral drugs used to treat AIDS or hepatitis might be effective against the SARS virus. Other researchers are investigating interferon, an antiviral protein produced by healthy cells exposed to a virus, to see whether it might be an effective treatment for SARS.

SARS is considered a rare disease because there were slightly more than 8,000 confirmed cases worldwide between 2002 and 2004, and none have been reported since April 2004. Public health doctors do not know whether SARS is likely to reemerge at some point in the future. In addition, some researchers are concerned about the possibility that the SARS virus might be used as an agent of bioterrorism.

SEE ALSO Common cold; Influenza; Pneumonia

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# Sexually Transmitted Diseases

Sexually transmitted diseases (STDs), also called venereal diseases, are contagious illnesses transmitted by vaginal or anal intercourse and by oral sex. Some STDs can also be transmitted by kissing, by an infected mother to her child during childbirth or breastfeeding, or by sharing needles used for intravenous drugs. Some doctors prefer the term "sexually transmitted infection" (STI) to STD because a person can be infected, and possibly infect others, without having obvious symptoms of a disease. Sexually transmitted diseases can be caused by a variety of disease agents. Some—such as AIDS, genital herpes, and human papillomavirus (HPV) infection—are caused by viruses. Others—including syphilis, gonorrhea, and chlamydia—are caused by bacteria. One type of STD, pubic lice or "crabs," is caused by a parasite.

Sexually transmitted diseases can cause long-term health problems if not promptly diagnosed and treated. Women may find themselves unable to have children. Such diseases can also cause blindness or other lifelong disabilities in children born to mothers with STDs. Some types of HPV infection increase a woman's risk of cervical cancer. In addition, any sexually transmitted disease is a risk factor for HIV infection and AIDS—a disease that still has no cure.

**SEE ALSO** AIDS; Chlamydia; Genital herpes; Gonorrhea; HPV infection; Lice infestation; Syphilis

# Sh

#### Also Known As

Shaken infant syndrome, SBS, abusive head trauma, pediatric traumatic brain injury

Genetic

Infection

Injury

Other

Multiple

Unknown

#### Cause

Violent shaking of an infant or toddler by an adolescent or adult

#### Symptoms

Brain damage, learning disabilities, mental retardation, blindness, paralysis, seizures, death

#### Duration

May be lifelong



# Shaken Baby Syndrome

#### Definition

Shaken baby syndrome (SBS) is a form of child abuse in which an adult forcefully shakes a baby or toddler by the arms, chest, or shoulders, causing the head to whiplash back and forth on the neck and leading to bleeding inside the skull from broken blood vessels. The syndrome was first identified by Dr. John Caffey (1895–1978), a pediatric radiologist, who published a landmark paper on it in 1972.

#### Description

Shaken baby syndrome is a type of traumatic brain injury that results when a baby or young child is grasped by the upper body and shaken back and forth by a teenager or adult. A baby has a relatively large and heavy head in comparison to the rest of its body, and it has weaker neck muscles than older children or adults. When the baby is shaken, or its head is struck against a wall or other hard surface, its brain moves back and forth inside the skull, causing tissue to bruise and small blood vessels to break. Blood can collect inside the skull and put pressure on the brain, leading to permanent brain damage, seizures, or death.

#### **Demographics**

The American Academy of Pediatrics (AAP) estimates that there are between 600 and 1,400 cases of SBS in the United States each year,
# Advice for Caregivers about SBS (source: The Arc)

There are three key words to remember when dealing with a crying or fussy baby: Stop. Calm down. Try again:

- Stop: Do not handle the baby if you are upset or angry. Place the child in a safe place like a crib or playpen.
- Calm down: Leave the room but stay close enough to hear the baby. Listen to calming music for a short time; then call a friend or the hotline below for support or advice. Another approach is to run the vacuum cleaner; the noise will drown out the sound of the crying; it also calms some babies. Keep in mind, too, that the baby may be crying from an earache, teething, or other illness as well as hunger or a wet diaper. If the baby cannot be soothed and keeps crying for a long time, it is best to call the doctor.
- Try again: After calming down, try again to help the baby.

Keep the number of the Childhelp National Child Abuse Hotline on the refrigerator or near the telephone: 1-800-4-A-CHILD (1-800-422-4453). The hotline is staffed twenty-four hours a day, 365 days a year. though it is possible that the true number is higher because some cases are misdiagnosed as the result of accidental falls or auto accidents. What is known is that shaken baby syndrome is the most common cause of mortality and long-term disability in infants and young children due to physical abuse. The syndrome has been reported in infants as young as five days and children as old as five years, but most victims are two years of age or younger.

SBS occurs in all racial groups in the United States but is more likely to be caused by males than by females. Adult males in their early twenties are the perpetrators in 65 to 90 percent of cases; most often they are the baby's father or the mother's boyfriend. Female perpetrators are more likely to be a teenage babysitter or nanny than the baby's mother. The usual trigger for the abuse is crying lasting for several hours or repeated diaper soiling, although in some cases involving men, the abuser is angry because he is jealous of the attention the baby receives from its mother.

## **Causes and Symptoms**

The cause of SBS is brain damage resulting from bleeding beneath the skull and bruising of brain tissue due to the brain's moving up against the inside of the skull during shaking. In some cases the brain is also damaged by loss of its oxygen supply.

The symptoms of severe SBS include:

- Bleeding into the retina of the eye
- Bleeding into the space between the brain and the layers of tissue that cover the brain
- Swelling of the head from fluid accumulating in the tissues of the brain
- Damage to the spinal cord and soft tissues of the neck

- Fractures of the ribs or other bones
- Convulsions
- Loss of consciousness

Babies who are less severely injured when shaken may have symptoms that are easy to confuse with the symptoms of flu:

- Vomiting or other flu-like symptoms without fever or diarrhea
- Crankiness and irritability over a period of time
- Poor feeding, loss of appetite
- Breathing problems
- Unusual drowsiness

#### Diagnosis

The doctor's greatest help in making a correct diagnosis of shaken baby syndrome is a description of what happened by the perpetrator or a witness. In many cases an abuser will tell the doctor that the child fell or was in a car accident, or that the abuser shook the baby trying to revive it. One important clue is that the injuries caused by SBS are usually much more severe than would be caused by a fall or other accidental head injury.

The doctor can tell that a shaken baby has a closed-head injury by taking imaging studies, usually a CT scan or an MRI. X-ray studies will reveal broken ribs or other bones. Bleeding into the retina of the eye can be detected by an ophthalmologist (doctor who specializes in eye disorders). In some cases, the doctor may order laboratory tests to rule out meningitis and other infectious diseases that can affect the brain and cause a seizure or coma.

## Treatment

Children with severe injuries from shaken baby syndrome require emergency treatment, usually brain surgery to relieve pressure on the brain and respiratory support to help them breathe. Treatment of the blindness, learning disorders, mental retardation, and other long-term consequences of SBS may last for the rest of the child's life. These children often need special education services, physical therapy, speech therapy, eye treatment, psychotherapy, and occupational therapy.

## Prognosis

SBS has a high mortality rate. It is estimated that a third of the babies who are abused in this way will die; another third will suffer severe permanent injuries; and the remaining third will recover.

#### Prevention

Dr. Caffey believed in the value of education to prevent at least some instances of SBS. While some abusers are people with a history of substance abuse or poor impulse control, others do not understand how much an angry adolescent or adult can harm a baby by shaking it. Various prevention strategies that are used include showing videos about SBS to new parents; encouraging pediatricians to discuss the stresses of childrearing with parents and teach them some ways to soothe a crying child; asking social workers to help identify families at risk of child abuse; instructing workers in day care centers and others who work with small children about the syndrome; and advising parents to screen babysitters or nannies very carefully before hiring them for child care responsibilities.

Specific tips for managing the stress of caring for a crying infant are described in the sidebar.

## The Future

It is difficult to tell whether the various preventive strategies for lowering the rate of shaken baby syndrome will have a significant effect in the years to come, as many abusive, addicted, or mentally disturbed people cannot be reached by educational measures. One hopeful development is that more doctors and nurses are aware of the syndrome and better able to diagnose it quickly.

SEE ALSO Child abuse; Whiplash

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## WORDS TO KNOW

**Closed-head injury:** An injury to the head in which the skull is not broken or penetrated.

- **Meningitis:** Inflammation of the protective membranes that cover the brain and spinal cord.
- **Radiologist:** A doctor who specializes in medical imaging techniques to diagnose or treat disease.
- **Retina:** The layer of light-sensitive tissue at the back of the eyeball.

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## Sickle Cell Anemia

## Definition

Sickle cell anemia is an inherited blood disorder caused by a mutation in a gene responsible for the production of hemoglobin, the protein found in red blood cells that carries oxygen from the lungs to other body organs and tissues. The disorder was first described in 1910 by James Herrick (1861–1954), a doctor in Chicago who was examining a young black dental student from the West Indies. Herrick described the cells seen in a smear of the patient's blood under the microscope as "thin, sickle-shaped and crescent-shaped red cells." The dental student died in 1916 at the young age of thirty-two.

A person must inherit the defective gene that produces hemoglobin S from both parents in order to develop sickle cell anemia. A person who inherits the gene from only one parent is said to have sickle cell trait. People with sickle cell trait do not have the symptoms of the disease but can pass on the disease if they have children with another carrier.

## Description

In sickle cell anemia, the red blood cells do not contain normal hemoglobin but a defective form of the protein called hemoglobin S. Whereas normal red blood cells are round, flexible, and able to move easily through the blood vessels, cells containing hemoglobin S can bend into crescent or sickle shapes and become sticky. They cause health problems for two reasons. First, blood cells containing hemoglobin S die much faster than normal red blood cells; this lowers their ability to carry enough oxygen to meet the body's needs and leads to such symptoms as lack of energy and lightheadedness or dizziness. Second, the abnormal red blood cells tend to form clumps that block blood vessels and cause organ damage.

When the sickle cells block blood vessels, they cause such symptoms as chest pain, damage to the spleen, liver, and kidneys, and stroke. Twothirds of all strokes in people with sickle cell anemia occur in children, with an average age of eight years. Another common complication is sickle cell crisis, a sudden severe attack of pain that affects the patient's chest, joints, abdomen, bones, or extremities.

Damage to the spleen caused by the abnormal blood cells makes patients with sickle cell anemia vulnerable to certain infections. Last, the effects of the sickle cells on the narrow blood vessels in the eyes lead to vision problems that include disorders of the retina and bleeding into the eye.

## **Demographics**

Geneticists think that the defective gene responsible for hemoglobin S originated independently in five different parts of Africa, including Cameroon and Senegal, and Saudi Arabia. The gene did not die out because, even though people with two copies of the defective gene did not usually live long enough to have children, those who had only the sickle cell trait were more resistant to one type of malaria. This advantage helped them to survive in parts of the world where malaria is

#### Also Known As

Sickle cell disease, hemoglobin SS disease, HbS disease

#### Cause

Mutation in a gene on chromosome 11

#### Symptoms

Fatigue, headache, pain attacks, swollen hands and feet, infections, vision problems

#### Duration

Lifelong



*Image of red blood cells, some misshapen as a result of sickle cell anemia.* DR. DAVID M. PHILLIPS / VISUALS UNLIMITED / GETTY IMAGES.

widespread. Sickle cell anemia is still more common in Africa, India, parts of Central and South America, the Caribbean islands, Saudi Arabia, and Turkey than in Europe or North America.

In the United States, sickle cell anemia primarily affects African Americans. About 80,000 people in the United States presently have sickle cell disease, which makes it the most common inherited blood disorder in North America. It occurs in one in every 375 live births among African Americans. According to the National Institutes of Health (NIH); 2 million Americans have sickle cell trait—one in twelve African Americans and one in every 100 Hispanics.

Males and females are equally likely to inherit either the trait or the disease.

## **Causes and Symptoms**

Sickle cell anemia is caused by a mutation in the hemoglobin-beta gene on chromosome 11.

The symptoms of sickle cell anemia do not usually appear in infants until they are about four months old. Severity differs from patient to patient.

• Anemia. Symptoms of sickle cell disease related to anemia include fatigue, pale skin, dizziness or feeling faint, cold hands or feet, and shortness of breath.

## A Great Jazz Musician

Miles Davis (1926–1991) was a jazz trumpeter and composer who was considered one of the most influential American musicians of the twentieth century. Davis helped to shape most of the major developments in jazz from the World War II period through his death in 1991. The son of a dentist in Alton, Illinois, Davis started to play the trumpet at age thirteen. He won a scholarship to the prestigious Julliard School of Music in New York in 1944, but quit to make his first professional recordings in 1945.

Davis was fortunate in suffering from a relatively mild form of sickle cell disease. He did, however, make his condition worse by becoming addicted to heroin and other drugs during his first visit to Paris in 1949. He had to take frequent breaks from his career in the 1960s and 1970s because of the impact of his drug abuse on the health of his bones, already weakened by sickle cell anemia. Davis eventually had to have a hip replacement in 1976. His death at the age of sixty-five was caused by a stroke combined with pneumonia, both complications of a sickle cell crisis.

- Sickle cell crises. These are episodes of severe pain caused by defective red blood cells blocking the flow of blood through the blood vessels that supply the chest, joints, bones, or abdomen. Crises occur suddenly and may last for anywhere from a few hours to several weeks. Some people have only a few crises during their lifetime while others may have a dozen or more every year.
- Vulnerability to infection. People with sickle cell anemia are more likely to get frequent infections because of damage to the spleen caused by the disease. Pneumonia, a lung infection, is the most common cause of death in children with sickle cell anemia.
- Jaundice. Jaundice is a condition in which the whites of the eyes and the skin have a yellowish discoloration because of liver problems.
- Hand/foot syndrome. This is often the first symptom of sickle cell anemia in babies. The child's hands and feet become swollen because the defective blood cells block the veins in the hands and feet and do not allow blood to return freely to the circulation.
- Eye disorders. Defective red blood cells blocking the tiny blood vessels in the eyes can

damage the retina, the light-sensitive layer of tissue at the back of the eyeball.

- Stunted growth and delayed puberty.
- Stroke. Although stroke is unusual in most children, about 11 percent of children with sickle cell disease suffer a stroke before they are twenty years old. Convulsions and partial paralysis often accompany stroke in this age group.

## Diagnosis

As of 2008, forty-nine states in the United States routinely screened newborns for sickle cell anemia through a blood test. A small amount of blood from the baby's finger or heel and sends is sent to a laboratory where it is analyzed for hemoglobin S. Older children and adults can also be tested for hemoglobin S with a blood test.

If the screening test is negative, no further testing is required. If the test results are positive for hemoglobin S, a second test is done to determine whether the person has sickle cell trait or the disease itself.

## Treatment

Treatment depends on the type and severity of the patient's symptoms. The only cure for sickle cell anemia is a bone marrow transplant (BMT). Only about 18 percent of children with the disease, however, have a suitable donor—usually a full sibling. The transplant procedure itself is complex, and 6 percent of children die during the procedure. As of 2008, BMT was usually performed only on children with a high risk of stroke, major risk of bleeding in the brain, severe visual disorders, and more than two sickle cell crises per year for several years.

Medications are the mainstay of treatment for patients with sickle cell anemia:

- Pain relievers. Depending on the severity of pain, patients with sickle cell anemia may be given prescription pain relievers during a sickle cell crisis. In some cases they may be taken to the hospital for intravenous fluids and strong narcotic painkillers.
- Antibiotics. Children diagnosed with sickle cell anemia are usually given penicillin from two months to five years of age to prevent infections. Adults with the disease may also be given antibiotics if they develop a bacterial infection.
- Hydroxyurea. Hydroxyurea is a drug developed to treat cancer that helps some people with sickle cell disease. It stimulates the production of fetal hemoglobin, a type of hemoglobin usually found only in newborns. It can reduce the need for blood transfusions and the frequency of sickle cell crises.
- Supplemental oxygen. Patients who are hospitalized with a severe crisis may be given supplemental oxygen to breathe to help raise the levels of oxygen in their blood and body tissues.

Blood transfusions lower the risk of stroke and relieve anemia in children with sickle cell anemia. Healthy red blood cells are removed from donated blood and infused into the veins of sickle cell patients. The risk of blood transfusions, however, is the buildup of iron in the patient's body, which can damage the liver and other organs. The Food and Drug Administration approved the drug Exjade in 2005, which removes excess iron from the blood.

Some lifestyle adjustments can help to lower the risk of sickle cell crises. Fevers, exposure to cold weather, becoming dehydrated, and using recreational drugs and alcohol have been found to trigger crises. Avoiding exposure to infections, drinking plenty of fluids, lowering emotional stress levels, and avoiding drugs and alcohol are all recommended for those with sickle cell anemia.

#### Prognosis

The prognosis for sickle cell anemia is still relatively poor. With the exception of children who benefit from bone marrow transplantation, most people with sickle cell anemia have shortened life expectancies. As recently as the 1990s, the average life span for patients with the disease was forty-two years for males and forty-eight years for females. As of 2008, about half of patients diagnosed with the disease lived into their early fifties.

## Prevention

The only way to prevent sickle cell disease is genetic counseling and testing. People can be tested for the sickle cell trait before starting a family and can talk to a counselor about their risk of having a child with sickle cell disease. It is also possible to test an unborn baby for sickle cell disease by a procedure called amniocentesis. The doctor uses a needle to withdraw a small sample of the fluid surrounding the baby in the mother's uterus. The cells in the sample are then tested for the defective hemoglobin S gene.

## The Future

There were several potential treatments for sickle cell anemia under investigation as of 2008. One is gene therapy. Experiments done in mice in the early 2000s have encouraged some researchers to think that it might be possible to remove some of the bone marrow cells in patients with sickle cell anemia, replace the defective hemoglobin-beta gene with a normal gene, and return the "corrected" cells to the patient's bone marrow.

## WORDS TO KNOW

**Anemia:** A condition in which a person's blood does not have enough volume, enough red blood cells, or enough hemoglobin in the cells to keep body tissues supplied with oxygen.

**Hemoglobin:** An iron-containing protein in red blood cells that carries oxygen from the lungs to the rest of the body.

Sickle cell crisis: Sudden onset of pain and organ damage in the chest, bones, abdomen, or joints caused by defective red blood cells blocking blood vessels.

Other treatments that are considered experimental are clotrimazole, an antifungal medication that appears to slow the production of sickle cells; and nitric oxide, a gas that helps to keep blood vessels open and reduce the stickiness of sickle cells.

SEE ALSO Malaria; Stroke; Thalassemia

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## SIDS See Sudden infant death syndrome.

## u Sjögren Syndrome

## Definition

Sjögren (pronounced SHOW-gren) syndrome, or SS, is an autoimmune disorder that primarily affects the body's tear glands and salivary glands. It is named for Henrik Samuel Sjögren (1899ndash;1986), the Swedish ophthalmologist (eye specialist) who described it in 1933. The disorder is also known as Gougerot disease or Gougerot-Sjögren syndrome, after Henri Gougerot (1881–1955), a French doctor who presented a case study of the disease in a French medical journal in 1925.

## Description

Sjögren syndrome is a disorder in which the body's immune system attacks the glands that produce saliva, tears, and (in women) vaginal fluid. It can also cause dryness in such other body tissues and organs as the kidneys, digestive tract, blood vessels, lung, liver, pancreas, and the central nervous system. Most patients also experience fatigue and pains in the joints.

Doctors classify cases of Sjögren syndrome as either primary, meaning that the disorder occurs by itself; or secondary, which means that the patient has another connective tissue disorder. About half of all cases of SS are primary and the other half are secondary. The four most common diseases affecting patients with secondary Sjögren syndrome are rheumatoid arthritis, lupus, scleroderma, and polymyositis (inflammation of the muscles).

## Demographics

SS is thought to affect between 0.1 percent and 3 percent of the population in all countries. According to the American College of Rheumatology (ACR), between 400,000 and 3.1 million adults in the United States suffer from the disorder. Members of all races appear to be equally affected. SS is primarily a disorder of women, however; females get SS

#### Also Known As

SS, Gougerot disease, Gougerot-Sjögren syndrome

#### Cause

Possibly a combination of genetic factors and an environmental trigger

#### Symptoms

Dry eyes, dry mouth, fatigue, vaginal dryness, joint swelling and stiffness

#### Duration

Years



*Salivary gland of a person with Sjögren syndrome.* © BIODISC/ VISUALS UNLIMITED / ALAMY.

nine times as frequently as males. It is unusual for adolescents or young adults to develop Sjögren syndrome; the average age of patients at onset is the late forties.

## **Causes and Symptoms**

The cause of Sjögren syndrome is not completely understood, but it is thought to be a combination of genetic factors, hormones, and environmental triggers, possibly infections. There is more than one gene that appears to be involved, but the connections between these genes and the onset of the disorder are not yet clear. Some researchers think that SS is triggered when people who are genetically susceptible are infected by a virus that has not yet been identified, and that the resultant infection causes the person's immune system to turn against the glands that produce tears and saliva. These glands include the tear glands near the eyes and the parotid glands below and in front of the ears.

The most noticeable symptoms of Sjögren syndrome are xerostomia, or dry mouth, and dry eyes. Patients with xerostomia may feel like their mouth is full of cotton or that they cannot easily talk or swallow. They are at increased risk of tooth decay because normal levels of saliva in the mouth help to protect against mouth infections and tooth decay. Patients with dry eyes may have a burning sensation in the eyes, blurred vision, or a gritty feeling beneath the eyelid. Drying of the eyes increases the risk of eye infections and damage to the cornea, the clear front part of the eye where light enters.

Other symptoms that patients may have include:

- Pain and stiffness in the joints and muscles
- Dry, itchy skin
- Rashes on the skin of the arms and legs
- Tingling sensations or numbness in the arms and legs
- Dryness in the vagina (in women)
- Chronic dry cough
- Fatigue or tiredness that is severe enough to interfere with daily life
- · Enlargement of the parotid glands

#### Diagnosis

The diagnosis of Sjögren syndrome is a long and complex process that may take years because the symptoms of the disorder are not specific to it; dry eyes and mouth, for example, can be caused by certain medications, by radiation therapy, or by anxiety disorders as well as by SS. Dry eyes can also be caused by disorders or factors other than SS. The National Institute of Arthritis and Musculoskeletal and Skin Diseases (NIAMS) found that patients who were eventually diagnosed with Sjögren syndrome received the diagnosis from primary care doctors, allergists, dentists, and cancer specialists as well as from rheumatologists, who are doctors who specialize in disorders of the muscles, joints, and connective tissue. Many rheumatologists think that the diagnosis of SS is often missed.

The diagnosis is usually based on a combination of the patient's symptom history and the results of laboratory tests and imaging studies. In taking the history, the doctor will ask about medications the patient may be taking and the amount of fluid that she or he drinks in an average day. In addition, the doctor will examine the patient's mouth for evidence of an abnormally low production of saliva. In many cases of SS, the patient's tongue will be dry enough to stick to the doctor's tongue depressor. Specific laboratory tests and imaging studies may include:

- Blood tests. These are done to check the blood cell count and also look for the presence of autoantibodies, which are proteins formed when to body's immune system attacks its own tissues.
- Schirmer tear test. This is a test in which the doctor measures the amount of tears produced by the patient's tear glands by placing a

small piece of filter paper underneath the lower eyelid for five minutes and then measuring its wetness with a ruler.

- Another eye test that may be performed involves staining the surface of the patient's eye with a dye called rose bengal. The dye will cling to areas of the cornea that have become dried out from SS. The doctor can then examine the patient's eyes through a slit lamp to determine how much the eye has been damaged by dryness.
- Sialogram. A sialogram is a special type of imaging study in which a radioactive dye is injected into the patient's parotid glands. The movement of the dye will help the doctor measure the flow of saliva from the gland into the patient's mouth.
- Lip biopsy. The doctor may take a small sample of lip tissue to check for the presence of inflammatory cells associated with SS.
- Urine test. A urine sample may be analyzed to see whether the patient's kidneys have been affected by SS.

## Treatment

There is no cure for Sjögren syndrome. Treatment for the disorder is aimed at relieving symptoms. Dry eyes can be treated by the application of artificial tears in the daytime and lubricating gels or ointments at night. The gels should not be used in the daytime because they can cause blurring of vision. Another drug that can be used is called Lacriserts. It comes in the form of small tablets that the patient places in the lower eyelid. When artificial tears are added, the Lacriserts tablet dissolves and forms a film over the patient's tears that traps moisture.

Patients with dry mouth can be given either Salagen or Evoxac. These are prescription medications that stimulate glands in the mouth to produce more saliva. Patients with SS must see their dentist on a regular basis to prevent tooth decay and eventual loss of teeth. In some cases the dentist may recommend fluoride treatment to provide additional protection for the teeth. Many patients find that taking small frequent sips of water or sugar-free lemon drops helps to relieve dryness in the mouth.

Patients with muscle or joint pains caused by Sjögren syndrome can take nonsteroidal anti-inflammatory drugs, or NSAIDs. These drugs include aspirin as well as such pain relievers as ibuprofen and naproxen, and are usually available over the counter. Steroid medications can also be given to patients with SS, but the long-term use of these drugs can cause serious side effects.

#### Sjögren Syndrome

## WORDS TO KNOW

- **Cornea:** The transparent front part of the eye where light enters the eye.
- **Lymphoma:** A type of cancer that affects the lymphatic system.
- **Parotid glands:** Glands that produce saliva, located on each side of the face below and in front of the ear.
- **Polymyositis:** Inflammation of the muscles that causes weakness and difficulty in moving or swallowing.

- **Rheumatologist:** A doctor who diagnoses and treats diseases of the muscles, joints, and connective tissue.
- **Scleroderma:** A disorder of connective tissue characterized by thickening and tightening of the skin as well as damage to internal organs.
- **Slit lamp:** An instrument that focuses light into a thin slit. It is used by eye doctors to examine eyes for a wide variety of disorders.

Xerostomia: The medical term for dry mouth.

## Prognosis

The prognosis of patients with Sjögren syndrome is generally good. About 5 percent of patients with SS eventually develop lymphoma, or cancer of the lymph nodes. Some patients also develop inflammatory disorders of the lungs, kidneys, or liver. The majority of patients, however, are able to manage the symptoms of the disease quite well and have a normal life expectancy. The most common complications of SS for the majority of patients are an increased risk of dental cavities and eye infections.

#### Prevention

There is no known way to prevent Sjögren syndrome because the causes of the disorder are not yet completely understood.

#### The Future

Sjögren syndrome is not likely to become more common in the general population because it is not inherited and is not contagious. Research in the early 2000s is focused on searching for the cause of the disorder, improving the speed and accuracy of diagnosis, and looking for potential cures.

SEE ALSO Lupus; Lymphoma; Rheumatoid arthritis; Tooth decay

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# m

## Skin Cancer

## Definition

Skin cancer is the uncontrolled growth of abnormal cells in the upper layer of the skin. It is the most common form of cancer among American adults. According to the American Cancer Society (ACS), more than a million cases of skin cancer are diagnosed each year in the United States.

## Description

Skin cancers develop in the epidermis, which is the outermost layer of the skin and contains several layers of cells within itself. In these layers there are several different types of cells. Skin cancers are classified by the type of epidermal cell that is affected:

• Basal cell carcinoma (BCC). Basal cells are cells in the lowermost layer of the epidermis. Basal cell carcinoma is the single most common type of skin cancer. These cancers appear on the head, neck, and hands as small fleshy bumps, nodules, or red patches.

Also Known As Carcinoma of the skin; melanoma

#### Cause

Genetic factors and exposure to ultraviolet radiation

#### Symptoms

Unexpected changes in the color or texture of the skin; growths on the skin that bleed, appear suddenly, or do not heal

**Duration** Months to years Tanning, from natural sunlight or from the use of tanning beds, can increase a person's risk of skin cancer. SHUTTERSTOCK.



They are slow-growing and rarely metastasize (spread to other parts of the body).

- Squamous cell carcinoma (SCC). Squamous cells are flat scale-like cells that form one or more layers in the middle layer of the epidermis. SCC is the second most common type of skin cancer. It may be preceded by precancerous patches of scaly skin called solar keratoses, which develop from overexposure to sunlight. Squamous cell carcinomas are most likely to appear on the ears, lips, tongue, mouth, or face. Unlike basal cell carcinomas, SCCs can invade deeper layers of tissue and spread to other parts of the body.
- Melanoma. Melanoma is the rarest type of skin cancer but also the deadliest. It develops in the melanocytes, which are cells in the lower part of the epidermis that produce a dark brownish-black pigment called melanin. Melanoma may appear suddenly as a dark patch on the skin or develop in or near a mole. It can spread rapidly to nearby lymph nodes and then to the liver, lungs, or brain.

## Demographics

Skin cancer affects adults far more often than children; basal cell carcinoma rarely occurs in people younger than forty years of age. The average age of people diagnosed with melanoma is fifty-three; however, it is the most common cancer in women aged twenty-five to twenty-nine years and is second only to breast cancer in women aged thirty to thirty-four years.

About one in five persons in the United States will develop skin cancer in his or her lifetime. Most skin cancers—96 percent—are either basal cell carcinomas or squamous cell carcinomas. Though they rarely cause death, these two types of skin cancer cost the United States over \$1.5 billion for treatment each year.

Melanoma accounts for only 4 percent of skin cancers in the United States, but it is responsible for 75 percent of deaths from skin cancer. About 8,500 Americans die each year from melanoma, 5,500 men and 3,000 women. Although melanoma is more common in women than men up to age forty, in adults over forty it is more common in men. In the United States, melanoma affects Caucasians twenty times more often than African Americans, and six times more often than Hispanics.

Risk factors for skin cancer include:

- Having fair skin that freckles or burns easily
- Living in areas that get high levels of ultraviolet radiation, such as mountainous regions or countries closer to the equator
- · Having a job that requires working outdoors during daylight hours
- A family history of skin cancer
- Having a disorder that affects the immune system, or a history of radiation therapy for cancer
- Having scars or burns on the skin (Fragile skin is more easily damaged by sun.)
- Exposure to certain chemicals in the environment, including arsenic and some types of weed killers
- History of blistering sunburns in childhood or adolescence
- Having a large number of moles
- Developing solar keratoses

## **Causes and Symptoms**

The basic cause of skin cancer is overexposure to the sun or to artificial sources of ultraviolet light like sunlamps or tanning booths. Ultraviolet light can damage the DNA, or genetic information, in skin cells, changing their genetic code and altering the function of those cells. Uncontrolled multiplication of these damaged skin cells can result in cancer.

Genetic factors are also involved. Skin color and sensitivity to the sun are inherited characteristics. In addition, melanoma is known to run in some families. Researchers have identified mutations in genes on chromosomes 1, 9, and 12 as linked to familial melanoma.

The symptoms of basal cell and squamous cell carcinomas include:

- Waxy or pearly bumps on the face, ears, or neck
- Flat brownish patches on the chest or back
- Firm red lumps or nodules on the face, lips, ears, neck, hands or arms
- Flat patches or lesions with scaly, crusted surfaces on the face, ears, neck, hands or arms

The signs of melanoma are sometimes called the ABCDs. A mole developing into melanoma may show:

- Asymmetry. Half of the mole looks different from the other half in size, shape, or color.
- Border irregularity. The mole has a ragged or poorly defined border.
- Color. The color is not uniform; the mole has dashes of blue, red, or white mixed in with patches of brown or black.
- Diameter. The mole is larger than the end of a pencil eraser (about 6 millimeters).

Melanoma may also appear as skin growths that bleed easily or do not heal.

#### Diagnosis

About 80 percent of skin cancers are found by patients who notice suspicious changes in their skin and go to their doctor. To diagnose skin cancer, the doctor will take a small sample of the abnormal skin to be sent to a laboratory for analysis under a microscope. This procedure is called a biopsy. The doctor may remove the entire growth, part of the growth, or shave a layer of cells from its surface. Biopsies are done under local anesthesia.

#### Treatment

Surgery is the most common treatment for skin cancer and solar keratoses. If the entire growth was removed for a biopsy, no further treatment may be needed. In some cases the cancerous areas can be removed with topical (applied directly to the skin) medications. Other treatment options include:

- Cryotherapy. This approach involves freezing the abnormal growths with liquid nitrogen.
- Standard surgery. The doctor cuts out the entire cancer and surrounding tissue. If the cancer is on the face, plastic surgery may be required later to restore the patient's appearance.
- Electrodessication. This is a form of treatment that involves desiccating, or drying out, skin cancers with an electric needle. It is usually done to treat small or shallow basal cell carcinomas.
- Laser surgery. This type of surgery is often done to treat cancers on the lips or cancers in the upper layers of the skin.
- Mohs surgery. Named for the surgeon who developed the technique in the late 1930s, Mohs surgery involves removing very thin layers of cancerous tissue and examining each layer under the microscope until all the cancer has been removed while leaving nearby healthy tissue unaffected. It is generally used for basal cell and squamous cell carcinomas.
- Radiation therapy and chemotherapy. These are used to treat metastatic melanoma and large BCCs and SCCs when surgery is not an option.

#### Prognosis

The prognosis of skin cancer depends on a number of factors: the patient's skin type, the type of cancer involved, the length of time before diagnosis and treatment, and the patient's overall health. In general, skin cancer is highly curable. Basal cell carcinoma and squamous cell carcinoma do not spread to other parts of the body as readily as melanoma; they have a 95 percent cure rate when treated early.

Melanoma, however, can metastasize to other parts of the body and is potentially deadly. Patients diagnosed and treated before their melanoma spreads to the lymph nodes have a five-year survival rate of 91 percent; however, those whose melanoma has spread to the lungs or liver have a five-year survival rate of only 7–10 percent, with an average life expectancy of six to nine months.

#### Prevention

People cannot change their skin type, but they can lower their risk of skin cancer by taking the following precautions against sun exposure:

- Avoid the use of tanning booths and sun lamps.
- Stay out of the sun between 10 A.M. and 4 P.M.
- Use a sunscreen with a sun protection factor (SPF) of 15 or higher every day. People with very fair skin should use a product with an SPF of 30 or higher.
- Apply sunscreen over the entire body thirty minutes before going outside, and reapply the product every two hours.
- Use a lip balm that contains sunscreen.
- Wear clothing that covers as much of the body as possible, including a broad-brimmed hat and sunglasses to protect the eyes.
- Keep infants under six months out of the sun altogether, and using sunscreen on infants older than six months.

Another important form of preventive care is regular self-examination of one's skin. The American Academy of Dermatology (AAD) outlines the steps:

- A person should first become familiar with his or her birthmarks, moles, freckles, and other skin blemishes in order to spot new growths or suspicious changes.
- Use a well-lit private room with a full-length mirror; take along a handheld mirror in order to see the back, buttocks, and other parts of the body that require a second mirror.
- It is important to check all parts of the body, not just those exposed to sunlight. Begin with the upper body, front and back; then the arms. Women should look underneath their breasts.
- Sitting in front of the mirror, examine the legs, genitals, soles of the feet, and the skin between the toes.
- Examine the back of the neck and scalp using the handheld mirror. Part the hair at intervals to check the entire scalp.

People with an increased risk of skin cancer should see a dermatologist regularly and also ask their primary care doctor to look for changes in their skin during checkups.

## The Future

Skin cancer is a major concern to doctors around the world because the rates of all types of skin cancer are increasing worldwide. These increases are partly the result of increased longevity, as the risk of skin cancer rises in

## WORDS TO KNOW

- **Carcinoma:** The medical term for any type of cancer that arises from the skin or from the tissues that line body cavities.
- **Cryotherapy:** The use of extreme cold to destroy cancerous tumors or other diseased tissue. It is also called cryosurgery.
- **Dermatologist:** A doctor who specializes in diagnosing and treating skin disorders.
- Epidermis: The outermost layer of the skin.
- Melanin: A brownish-black skin pigment.
- Melanoma: The most serious form of skin cancer.

- **Metastasis (plural, metastases):** A secondary tumor caused by the spread of cancer from its primary location to another part of the body.
- **Mohs surgery:** A technique for removing skin cancers in very thin layers one at a time in order to minimize damage to healthy skin.
- **Solar keratoses (singular, keratosis):** Rough scaly patches that appear on sun-damaged skin. They are considered precancerous.
- **Topical:** Referring to any medication that is applied to the skin or the outside of the body.

people over forty and even higher in people over sixty-five. Another factor is the ongoing popularity of tanning, whether in the sun or under artificial sunlamps. Melanoma in particular has increased around the world since the 1990s, with the highest rates as of 2008 in Australia and New Zealand.

SEE ALSO Dermatitis; Sunburn; Xeroderma pigmentosum

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# **m** Sleep Apnea

## Definition

Sleep apnea is a sleep disorder in which a person's breathing stops and starts over and over in the course of their sleep time. There are three basic forms of sleep apnea: obstructive sleep apnea (OSA), in which the interruptions in breathing are caused by a physical blockage of the airway, usually loosened tissues in the back of the throat; central sleep apnea (CSA), which results from irregular signals from the part of the brain that controls breathing; and complex (or mixed) sleep apnea, which is a combination of obstructive sleep apnea and central sleep apnea. Obstructive sleep apnea is the most common type, accounting for about 90 percent of cases.

## Description

Sleep apnea is a sleep disorder characterized by interruptions in breathing in which the person misses one or more breaths over an interval of ten seconds or longer; these episodes of interrupted breathing occur repeatedly during sleep. Some people have as many as several hundred episodes of apnea during their sleep time.

In normal sleep, the muscles at the back of throat relax, but remain open far enough to allow air to pass through the airway into the lungs. In obstructive sleep apnea, the muscles in the back of the person's throat are too relaxed and allow the tongue, the tonsils, the soft palate, or the

Also Known As Sleep apnea syndrome

#### Cause

Periodic pauses in breathing during sleep

#### Symptoms

Snoring, daytime tiredness, morning headaches, trouble concentrating, changes in mood or behavior

#### Duration

Months to years unless treated or corrected

#### Sleep Apnea



Patient wearing a continuous positive airway pressure device used to diagnose and monitor patients with sleep apnea. © MEDICAL-ON-LINE / ALAMY.

uvula—a triangular flap of tissue at the back of the soft palate—to slip backward or downward and block the airway.

In some people, airway blockage may be the end result of certain physical features, such as an unusually narrow airway, a large neck, or a recessed chin. When the airway closes, the person typically begins to snore or gasp for breath. As the oxygen supply in the person's blood drops, the brain gives a signal that arouses the person from sleep in order to start breathing again. The person usually does not remember waking up in order to breathe; however, their sleep is not restful because of the frequent interruptions and the person feels tired or sleepy the next day.

Central sleep apnea is unusual in people who are otherwise healthy. It is most likely to occur in people with heart disease, stroke, or damage to the brain stem—the part of the brain that controls breathing. Central sleep apnea occurs when the brain fails to transmit its normal signals to the breathing muscles. The affected person may awaken feeling short of breath or have a difficult time falling or staying asleep. As with obstructive sleep apnea, central sleep apnea can result in snoring and daytime sleepiness.

Sleep apnea is a potentially severe long-term health problem. People who are not treated for sleep apnea have a 30 percent higher risk of heart attack or premature death because the drop in the oxygen level of the blood that occurs when breathing stops temporarily can eventually damage the heart. Sleep apnea also increases a person's risk of stroke and high blood pressure. In addition, a study conducted in 2007 reported that the brains of people with obstructive sleep apnea show evidence of tissue shrinkage in the parts of the brain that store memory. These findings help to explain why sleep apnea can lead to memory loss.

Sleep apnea can endanger the well-being of others as well as the patients themselves. Daytime sleepiness is a risk factor for motor vehicle accidents; one study showed that people with sleep apnea are three times more likely to be involved in accidents than people who do not have the disorder.

#### Demographics

Sleep apnea is a common sleep disorder in the general American population. According to the National Institutes of Health (NIH), as many as eighteen million adults suffer from sleep apnea, with as many as ten million more who have the condition but have not been diagnosed.

Several risk factors for sleep apnea have been identified:

- Age. Middle-aged adults are more likely to have sleep apnea than children or adolescents. About 4 percent of middle-aged men and 2 percent of middle-aged women in the United States have been diagnosed with sleep apnea, while about one person in ten over the age of sixty-five has the disorder.
- Sex. Men are twice as likely as women to develop sleep apnea
- Race and ethnicity. African Americans, Pacific Islanders, and Hispanics are at greater risk of sleep apnea than Caucasians, Asian Americans, or Native Americans.
- Weight. Obesity is a major risk factor in sleep apnea.
- Physical features. A large neck, narrow airway, small mouth or nose, enlarged tonsils, or a receding chin can all increase a person's risk of sleep apnea.
- Lifestyle. Drinking alcohol and smoking increase the risk of sleep apnea.
- Heredity. Sleep apnea is known to run in families, although no specific genes associated with sleep apnea have been identified as of 2008.
- Medications. Certain types of prescription drugs, particularly antidepressants, tranquilizers, and sleeping pills, increase the risk of sleep apnea.

• Allergies. Hay fever and other allergies that cause swelling of the tissues in the nose and throat increase a person's risk of sleep apnea.

## **Causes and Symptoms**

Sleep apnea has a number of possible causes ranging from congenital abnormalities in the shape of the nasal passages and airway to medical disorders or lifestyle choices that contribute to blockage of the upper airway during sleep.

In addition to daytime sleepiness, irritability, and difficulty concentrating, symptoms of sleep apnea may include:

- Loud snoring
- Waking up abruptly and feeling short of breath
- Waking with a dry mouth or sore throat
- Having a headache in the morning
- Difficulty staying asleep
- High blood pressure
- Swelling of the legs
- · Poor judgment and memory loss

#### Diagnosis

The diagnosis of sleep apnea is often missed, particularly in those who live alone, because people with the disorder often do not remember being aroused from sleep by the need to start breathing again. Those who share a bedroom with another family member, however, may consult their doctor when the other person complains about snoring or notices that the person with sleep apnea sometimes stops breathing completely. The doctor will take the patient's history, including a medication history, allergy history, and a family history of sleep apnea. The patient may be referred to a dentist or oral surgeon for detailed evaluation of their throat, mouth, and airway if appropriate.

The patient may also be referred to a sleep laboratory or sleep disorder center for polysomnography. A polysomnograph is a machine that measures the patient's heart, lung, and brain activity during sleep, along with their breathing patterns, arm and leg movements, and blood oxygen levels. There are also portable machines that will measure the patient's blood oxygen level, airflow, and breathing patterns during sleep. In some circumstances the doctor may allow the patient to use one of these simpler machines at home to help diagnose sleep apnea.

#### Treatment

The treatment of sleep apnea depends partly on its cause or causes. Changes in lifestyle—particularly losing weight, quitting smoking, and cutting back on alcohol intake—may be enough to control obstructive sleep apnea in some people. According to the American Association for Respiratory Care, losing as little as 10 percent of body weight can reduce the number of times a person with obstructive sleep apnea stops breathing during sleep. If the sleep apnea is related to prescription medications, the patient's doctor may be able to adjust the dosages or substitute different drugs. Nasal sprays or allergy medications may relieve sleep apnea caused by swollen nasal tissues.

Changes in sleeping position may also help. The NIH recommends sleeping on the side rather than the back to help keep the throat open during sleep. Special pillows that prevent the person from turning onto the back are available.

Another treatment that works well for some patients with mild to moderate sleep apnea is an oral appliance fitted by a dentist for use at night. These devices are designed to keep the throat open at night by bringing the jaw forward. The dentist will check the appliance periodically to make sure that it fits correctly and that the patient's symptoms are improving.

In children or adults with enlarged tonsils, a simple tonsillectomy may cure the sleep apnea. There are also surgical procedures that can be done to correct problems in the nasal passages, such as irregularly shaped passageways or benign growths of tissue called polyps. Severe obstructive sleep apnea, however, may involve one or more of the following types of surgery:

- Surgery to remove or shrink part of the uvula or the soft palate. This type of procedure is usually done in a hospital under general anesthesia.
- Relocation or repositioning of the upper and lower jaws. This procedure is usually done by a team that includes an oral surgeon and an orthodontist. Bringing the upper and lower jaws forward enlarges the space behind the soft palate and reduces the risk of airway obstruction during sleep.
- Tracheostomy. This procedure is usually performed only when the person's sleep apnea is life-threatening and other treatments have failed. The surgeon cuts a hole in the front of the neck and inserts

a metal or plastic tube to provide the patient with an artificial airway.

Patients with moderate or severe sleep apnea may benefit from continuous positive airway pressure, or CPAP. CPAP is a machine that delivers air through a mask into the patient's nose during sleep. Positive pressure means that the air delivered by the machine is under slightly greater pressure than the air in the room, and this additional pressure keeps the airway open. CPAP machines have adjustable pressure settings, and the mask can also usually be adjusted for the patient's comfort.

Patients with central sleep apnea often benefit from having the underlying medical condition treated, particularly if heart failure is involved. Supplemental oxygen therapy and CPAP are also used to treat central sleep apnea.

## Prognosis

The prognosis of sleep apnea depends on its cause and its severity. People with mild sleep apnea usually do very well with lifestyle adjustments, allergy treatments, or changes in medications. CPAP is an effective treatment for moderate to severe sleep apnea; however, many patients find its side effects (dry nose, irritated facial skin, and headache) bothersome and stop using it. For those patients, surgery is often an effective alternative. Most patients, even those with severe sleep apnea, find that some form of treatment for this disorder can significantly improve their quality of life; it may simply take time to find the most effective treatment for a specific individual.

## Prevention

Sleep apnea related to a family history of the disorder or the physical characteristics of a person's throat and facial structure cannot be prevented, only corrected. Keeping one's weight at a healthful level for one's sex, age, and height, drinking only in moderation, and quitting smoking (or never starting to smoke), however, can lower an individual's risk of developing sleep apnea in middle age.

## The Future

Sleep apnea is a condition that is presently attracting many researchers in the fields of psychology and neurology as well as sleep medicine. As of 2008, the National Institutes of Health was conducting 134 different

## WORDS TO KNOW

Apnea: Temporary stopping of breathing.

Congenital: Present at birth.

**Polysomnograph:** A machine used in a sleep laboratory to monitor chest movement, air flow,

brain waves, heart rhythm, and other data relevant to diagnosing sleep disorders.

**Uvula:** A triangular piece of soft tissue located at the back of the soft palate.

studies on sleep apnea, ranging from evaluations of surgical treatments and mouth appliances to new drugs and improvements in CPAP.

**SEE ALSO** Allergies; Childhood obesity; Heart failure; Obesity; Smoking; Sudden infant death syndrome

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## **m** Sleep Disorders

Sleep disorders are a group of health problems that include difficulty falling or staying asleep, falling asleep at inappropriate times, excessive total sleep time, or abnormal behaviors associated with sleep. Doctors generally classify sleep disorders into three broad categories: those that involve either sleeping too much or not getting enough sleep; those that involve sleepwalking, nightmares, and other forms of arousal from sleep; and those related to medical or psychiatric conditions that affect sleep.

The first category—disturbances in the amount of sleep—include narcolepsy, sleep apnea, and restless legs syndrome. Another type of sleep disorder in this category is jet lag, which is caused by changes in the person's normal sleep/wake cycle resulting from travel across several time zones. Older adults sometimes have trouble getting enough sleep because their sleep/wake cycle does not work as well as it did when they were younger.

Medical and psychiatric conditions that are associated with sleep disorders include alcoholism, depression, panic disorder, schizophrenia, and posttraumatic stress disorder.

Most adults need about eight hours of sleep to feel fully alert during their waking hours. Teenagers need even more—8.5–9 hours of sleep per night. Getting help for a sleep disorder is important because lost sleep can affect physical coordination (including safe driving) as well as emotions and the ability to pay attention and think clearly.

**SEE ALSO** Alcoholism; Depression; Narcolepsy; Panic disorder; Posttraumatic stress disorder; Restless legs syndrome; Schizophrenia; Sleep apnea



## Smallpox

## Definition

Smallpox is an infectious disease caused by the variola virus. Researchers think that it originated in rats or mice somewhere in Africa and was

transmitted to humans around 10,000 BCE. The disease presently affects only humans and cannot be acquired from any animal.

There are two types of smallpox, variola major and variola minor. Variola major is the more severe of the two, accounting for about 90 percent of cases; it has a mortality rate of 30–35 percent. Variola minor, sometimes nicknamed white pox, cotton pox, or Cuban itch, is a milder disease that kills less than 1 percent of those infected. Each type of smallpox makes a person who survives it immune to both forms of the disease.

## Description

Smallpox is one of the most destructive diseases known to humankind. It was first called smallpox in the fifteenth century to distinguish it from syphilis, which was then called the Great Pox. About 400,000 people died each year from smallpox in Europe alone for most of the eighteenth century. During the twentieth century between 300 and 500 million people around the world died from smallpox. The mortality rate rose as high as 60 percent in some countries, with 80 percent of infected children dying from the disease.

Smallpox was gradually eliminated following the development of an effective vaccine against it in 1796. Edward Jenner (1749–1826), an English doctor, had noticed that milkmaids rarely got smallpox and theorized that they were protected by exposure to cowpox, a disease similar to smallpox but much less deadly. Jenner tested his theory by inoculating an eight-year-old boy with pus taken from a milkmaid's cowpox blisters and showing that the boy was immune to smallpox. Jenner's early vaccine was refined and introduced in the developed countries during the nineteenth century. The term vaccination was derived from the French word for cow related to the original source of the immunization virus. By 1897, smallpox had been virtually eliminated in the United States; the last case of smallpox in North America was reported in 1949.

In the 1960s, the World Health Organization (WHO) led a worldwide effort to eradicate smallpox. A team of scientists led by Donald Henderson (1928–) was formed to vaccinate people in developing countries. The last case of naturally occurring smallpox was diagnosed in Somalia in 1977. In 1980 WHO declared that smallpox had been eradicated. As of 2008 two laboratories, one at the Centers for Disease Control and Prevention (CDC) and the other in Russia, maintained small stocks of the smallpox virus for research purposes.

#### Also Known As

Variola major, variola minor

#### Cause

Virus

#### Symptoms

High fever; rash that becomes pus-filled sores; vomiting; severe headache; delirium

#### Duration

Two to three weeks

#### Smallpox



*Smallpox sores on the hand and arm.* © MEDICAL-ON-LINE / ALAMY.

The disease itself is characterized by a prodrome, or period of warning symptoms, characterized by high fever (101–104°F [38.3°C–40°C]) and flu-like symptoms that include severe headache and fatigue. The prodrome lasts between two and five days and is followed by red spots on the face and in the mouth, developing into open sores that spread the virus within the mouth and throat. About a day later, a rash spreads over the body that turns into papules (raised pimples) within three days. Unlike chickenpox, the skin lesions of smallpox are all in the same stage of development at the same time. The papules fill with a pus-colored fluid and may feel as if a hard, small, marble bead is present under the skin. The papules gradually form scabs that fall off the body, often leaving scar tissue behind. In fatal cases, death occurs between ten and sixteen days after the first symptoms.

## Demographics

Smallpox is equally likely to infect people of any race or age group and people of either sex during an epidemic. The chief difference is the mortality rate.

People who are at increased risk of dying from smallpox if they become infected include:

- Pregnant women
- Young children
- Elderly adults
- Anyone who has never been vaccinated against smallpox

#### **Causes and Symptoms**

The cause of smallpox is the variola virus, which usually enters the body through the mouth and respiratory tract. In most cases the virus is usually transmitted from person to person by close contact (within 6 feet [2 meters]); smallpox is not as easily transmitted as measles or influenza. The virus has an incubation period of seven to seventeen days before the prodrome occurs. Once in the person's mouth or throat, the smallpox virus multiplies rapidly in nearby lymph nodes and then enters the bloodstream. It is carried to the spleen, bone marrow, liver, and kidneys. People who have been infected with the virus can transmit it to others late during the prodrome; however, they are most contagious when the rash appears.

In addition to high fever and the characteristic rash, the symptoms of smallpox may include:

- Severe headache
- Fatigue
- Muscle aches and pains
- Severe back pain
- Vomiting, diarrhea, or both
- Delirium
- In some cases, heavy bleeding in the internal organs

Patients who survive smallpox often have complications:

- Permanent scars on the face. Called pock marks, these scars occur in about 80 percent of people who survive the disease. Queen Elizabeth I of England and the Russian dictator Josef Stalin were scarred by the disease; the queen wore heavy makeup in later life to try to hide her pock marks.
- Eye infections. About 20 percent of survivors develop conjunctivitis; a few become permanently blind.
- Infection of the bone. Between 2 and 20 percent of children who get smallpox develop an infection of the bone or bone marrow known as osteomyelitis.

#### Diagnosis

The most important aspect of diagnosing smallpox is distinguishing it from chickenpox. This is particularly important since most doctors in developed countries have never seen a case of smallpox. There are three major differences between chickenpox and smallpox:

- The skin lesions of chickenpox occur mostly on the face and trunk rather than the face, arms and legs. They are also shallower and do not feel as if there is a hard object inside them.
- Chickenpox lesions occur in waves or crops, so that the patient may have lesions at different stages of development at the same time. In smallpox, the lesions are all at the same stage of development at a given time.
- Chickenpox is contagious before the infected person feels sick, whereas smallpox is not contagious until the symptoms appear.

Diagnosis is based on a combination of the patient's history particularly a prodrome with fever two to four days before the rash appeared, and the appearance of the rash (lesions are hard to the touch, appear to be deep within the skin, and appeared first on the face, mouth, and forearms)—and a laboratory test. To test for the virus, the doctor will swab the patient's throat or take a smear from one of the skin lesions. The sample will be sent to a state health department laboratory for examination under an electron microscope or tested by a technique called polymerase chain reaction or PCR. Under a microscope, a cell infected with the smallpox virus will have pink clumps of the virus known as Guarnieri bodies, named for the Italian doctor who discovered them. PCR is a technique that identifies the virus's DNA.

Accurate diagnosis of a suspected case of smallpox is critical because it would be considered a major public health emergency. In the United States, the Centers for Disease Control and Prevention (CDC) and the United States Army Medical Research Institute of Infectious Diseases (USAMRIID) must be consulted and public health officials notified.

#### Treatment

There is no medication that can be given to cure smallpox; the goal of treatment is to keep the patient alive and isolated from others until he or she is no longer contagious. Once the patient has been taken to the hospital, the mainstays of treatment are supportive care and antibiotics to prevent bacterial infections from developing. Vaccination with smallpox vaccine can lessen the severity of the infection if given within four days of infection. However, since the incubation period of the disease is longer than four days, vaccination is primarily useful in protecting health care workers and others who may have been in contact with the person.

The most important treatment measure is isolating the patient in order to prevent the disease from spreading further. The procedure that it usually followed is called ring vaccination. Everyone who has been in contact with the patient within the previous seventeen days, including hospital staff, is placed in quarantine and given smallpox vaccine. In addition, local, state, and federal health authorities are notified at once.

## Prognosis

The prognosis for recovery from smallpox depends on the form of the disease and the patient's age and general health. Variola major has a death rate around 30 to 35 percent in unvaccinated people, although the rate rises to 60 percent in unvaccinated pregnant women. Variola minor has a mortality rate of about 1 percent. There are rare forms of variola major called hemorrhagic smallpox and malignant smallpox, both of which have fatality rates above 99 percent.

#### Prevention

Smallpox can be prevented by vaccination. Although compulsory vaccination for the general population was discontinued in 1980 and for military personnel in 1989, vaccination against smallpox was reintroduced after the terrorist attacks of 9/11 for health care workers and members of the armed forces. Some public health officials are concerned about the possibility of a major smallpox epidemic in the event of a laboratory accident or act of bioterrorism, as the smallpox vaccine is thought to be effective for only ten years. In addition to the fact that most people in the United States have not been vaccinated since 1980, an estimated 42 percent of the population has never been vaccinated.

Apart from vaccination, the only preventive measures that are even partly effective are isolation and quarantine of persons thought to be infected with smallpox.

## The Future

The major concern for the future regarding smallpox is the possibility of large-scale epidemics in the event of accidents or terrorist attacks. Smallpox would be a deadly biological weapon because only ten to one hundred particles of the virus are needed to infect a person. In 1978, the

## WORDS TO KNOW

- **Bioterrorism:** The use of disease agents to frighten or attack civilians.
- **Delirium:** A suddenly developing mental disturbance characterized by confused thinking, difficulty focusing attention, and disorientation.

**Eradication:** The complete elimination of a disease.

Lesion: A general term for any skin injury.

**Papule:** A small cone-shaped pimple or elevation of the skin.

- **Prodrome:** A group of warning signs or symptoms that appear before the onset of a disease.
- **Quarantine:** The practice of isolating people with a contagious disease for a period of time to prevent the spread of the disease.

year after the last reported case of naturally occurring smallpox, two people in England died of smallpox through a laboratory accident. As a result of the accident, all known stocks of the smallpox virus were either destroyed or sent to two laboratories, one at the CDC and the other in Russia. The remaining supplies of the virus were originally scheduled to be destroyed in 1995, but the WHO decided in 2002 to preserve the virus for development of new vaccines, diagnostic tests, and drugs that might be effective in treating smallpox.

SEE ALSO Anthrax; Chickenpox; Conjunctivitis; Plague

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## **Smoke Inhalation**

### Definition

Smoke inhalation (SI) is a health problem and potential emergency caused by exposure to the heat and smoke of a fire; to the products of cigarette smoke; or to other sources of outdoor or indoor air pollution.

## Description

Smoke inhalation occurs when a person is forced to breathe in (inhale) the toxic gases and particles carried in smoke from a fire, volcanic eruption, or industrial process. Smoke can arise from natural as well as human causes, ranging from lightning strikes, wildfires, and volcanic eruptions to house or building fires, fires following railroad or aviation disasters, pipeline or factory explosions, arson and other criminal acts, and traffic exhaust. Some people also choose to inhale smoke, as when they light up a tobacco or marijuana cigarette.

Smoke inhalation damages the respiratory system in one or more of three ways. First, the hot air from a fire can damage the mouth and upper

#### Also Known As

Smoke-related injury, SI, inhalation injury

#### Cause

Heat, smoke, and other toxic products of a fire

#### Symptoms

Coughing, scratchy throat, difficulty breathing, headaches, chest pain, nausea, confusion

#### Duration

Seconds to hours



Mother and her baby in Kenya, who suffer from smoke inhalation from their cooking fire. ADAM HART-DAVIS / PHOTO RESEARCHERS, INC.

airway. This thermal injury can be even worse if the hot air is accompanied by steam. Second, a fire can starve the body of needed oxygen, either because the fire is using up the oxygen in a closed space, or because the fire is producing carbon monoxide and other gases that interfere with the blood's ability to transport oxygen to the brain and heart. Third, smoke often contains chemicals that irritate the tissues of the lungs, causing damage that ranges from triggering the body's immune system to complete direct destruction of lung tissue.

## **Demographics**

Smoke inhalation, rather than burn injuries, is responsible for the majority of fire-related deaths in the United States. Between 50 and 80 percent of deaths related to house fires are caused by SI rather than burns. Fires are the third leading cause of accidental deaths in all age groups in the United States. Arson is the single most common cause of house fires (26 percent), followed by faulty electrical wiring (17 percent) and faulty heating systems (16 percent).

Some persons are at greater risk of serious injury from smoke inhalation, including:

- Firefighters and emergency workers.
- People in occupations that involve making or transporting hazardous materials.

## Smoke Inhalation and Personal Safety

Smoke inhalation, or SI, can be a serious threat to health. The Centers for Disease Control and Prevention (CDC) recommend the following steps in case of exposure to smoke in a fire or other emergency:

- Get away from the source of the smoke as quickly as possible. If in a building, avoid smoke-filled hallways or stairwells. If there are no other ways out of the building, stay as low to the floor as possible and crawl under the smoke toward the closest exit.
- Once in an area with fresh air, rest for a few moments and take several slow deep breaths.
- Do not return to a burning home or other building until the fire has been completely extinguished and all the smoke has cleared.
- Call the doctor if someone exposed to smoke feels feverish or develops a high temperature.
- Call for emergency help *at once* if the person is wheezing, having trouble breathing, is vomiting, has burns on the face, or seems confused or unusually sleepy. In addition, do not let the injured person try to drive himself or herself to the hospital.

- Children, who are susceptible to airway damage because their airways are still developing and they breathe more air per pound of body weight than adults.
- Elderly persons.
- People with asthma, bronchitis, emphysema, or other disorders that affect the lungs.
- People who are heavy smokers.
- People who are under the influence of drugs or alcohol.
- Persons who are physically disabled.

## **Causes and Symptoms**

Smoke inhalation causes injury or death by a combination of heat damage to the tissues of the mouth and upper throat; oxygen starvation of body tissues; and chemical damage to the tissues of the lungs.

Some fires are more dangerous than others in terms of the inhalation injuries they can cause. There are several factors that affect the potential deadliness of smoke from a fire:

- Temperature of the fire. The hotter the fire, the greater the thermal injury to the upper airway.
- Location of the fire. Fires inside closed spaces, such as houses or other buildings, aircraft fuselages, railroad cars, etc. use up the oxygen that people trapped inside the space need to breathe.
- Materials being burned. Plastics, silk, and wool all release cyanide gas when burned. This gas increases the risk of damage to the central nervous system.

The symptoms of smoke inhalation include:

• Cough. The patient will usually bring up mucus, which may be either clear or black (if it contains soot or smoke particles).

- Difficulty breathing, rapid breathing, or hoarse or noisy breathing.
- Reddened eyes.
- Abnormal skin color. Patients who are oxygen-starved may have pale or bluish skin. A cherry-red color may indicate carbon monoxide poisoning.
- Soot in the nostrils or upper throat.
- Headache.
- Nausea and vomiting.
- Changes in level of mental alertness or consciousness. Patients who have been severely affected by smoke inhalation may faint, have seizures, or go into a coma.

In some cases the symptoms of smoke inhalation do not appear until a day or two after the fire. People who have been exposed to smoke but seem healthy should be observed or monitored at home for at least forty-eight hours after the fire. The doctor should be called if the person develops a hoarse voice, chest pains, long periods of coughing, or mental confusion.

#### Diagnosis

The diagnosis of smoke inhalation is based on a combination of the patient's history (which will be obvious if he or she has left or been rescued from a fire) and imaging or laboratory studies. Emergency rescue personnel can check the patient at the scene for evidence of facial burns, soot in the airway, and other external signs of SI. They can also measure the patient's breathing, pulse, and level of consciousness.

After the patient has been taken to the hospital, he or she will usually be given a chest x-ray to check for lung damage; an electrocardiogram to make sure that the heart is functioning adequately; and blood tests to measure the amount of oxygen in the blood or the presence of chemical byproducts of smoke inhalation. The doctor may also use a bronchoscope (a flexible lighted tube that allows the passages into the lungs to be examined and have fluid removed) to look for damage to the respiratory system. In a few cases the patient may be given a CT scan to assess possible brain injury.

### Treatment

Treatment is based on the severity of the patient's injuries, as SI can range from minor irritation of the tissues lining the airway to an immediate threat to life. The most important emergency measure is keeping the airway open and supporting the patient's breathing. The patient will be given oxygen through a mask or a tube inserted down the throat. The patient may also be given brochodilators, which are medications that relax the tissues in the airway and help to open up the breathing passages. Patients who have inhaled large amounts of carbon monoxide may be put in a hyperbaric oxygen (HBO) chamber, a special room in which the patient is given pure oxygen at two to three times normal atmospheric pressure.

Treatment of SI may also include suctioning of excess fluid from the lungs by means of a bronchoscope. This instrument can be used for treatment as well as diagnosis of SI.

Patients with mild symptoms from SI are usually kept in the emergency room for observation after treatment for four to six hours. They are advised to return to the hospital at once if their symptoms return or worsen. Those who were exposed to fire in a closed space for longer than ten minutes, have coughed up black mucus, have facial burns, are coughing severely, or have difficulty swallowing are usually admitted directly to the hospital for further treatment.

#### Prognosis

The prognosis of recovery from smoke inhalation depends on a number of factors, including the patient's age, previous health, the length of time one was exposed to the smoke, and whether he or she was burned in addition to inhaling smoke. The mortality rate from smoke inhalation by itself is about 7 percent; however, the mortality rate is about 29 percent for patients who suffer burns as well as SI.

Some people may have chronic shortness of breath or permanent scarring of the lungs following smoke inhalation. These long-term problems are particularly likely to develop in people who smoked, or had asthma or other lung disorders before being exposed to smoke from a fire.

#### Prevention

The CDC recommends the following measures to reduce the risk of illness or death from smoke inhalation:

- Install fire, smoke, and carbon monoxide detectors in the home and check them regularly. The absence of a smoke detector increases the risk of death in a fire by about 60 percent.
- Follow local air quality reports; listen for news reports about smoke or outdoor air pollution.

## WORDS TO KNOW

- **Arson:** The intentional setting of a fire in a building or other property. Arson is a criminal act in the United States.
- **Bronchodilator:** A type of medication that relaxes the tissues of a congested or smoke-damaged airway, thus making it easier for the patient to breathe.
- **Bronchoscope:** A flexible lighted tube that can be inserted into the passages leading to the lungs for examination or treatment.
- **Hyperbaric oxygen (HBO):** Oxygen that is delivered to a patient in a special chamber at two to three times normal atmospheric pressure.
- **Inhalation:** The part of the breathing cycle in which a person takes in air from the outside.
- If the local air quality index indicates that people should stay indoors, indoor air should be kept as clean as possible. Keep doors and windows closed. In hot weather, run the air conditioner with the fresh-air intake closed.
- Do not add to indoor air pollution by smoking or by burning wood in fireplaces when outdoor air quality is poor.
- Make an escape plan for the home to be followed in case of fire. Practice the escape route with the family, including evacuating pets.

## The Future

The United States has one of the highest rates of fire fatalities in the developed world—about 2.3 deaths per 100,000 population. A majority of these deaths are due to SI rather than burns. In addition to installing and properly maintaining smoke detectors and alarms, better methods of screening patients for injury from smoke inhalation are needed. One ongoing difficulty with emergency treatment of fire victims is the present lack of tests that are sensitive enough to identify patients who develop delayed reactions to smoke inhalation.

**SEE ALSO** Asthma; Bronchitis; Carbon monoxide poisoning; Emphysema; Smoking

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## <mark>m</mark> Smoking

## Definition

Smoking refers to the burning of tobacco (or other plant, like marijuana) in a cigarette, cigar, or pipe in order to inhale the smoke. Smoking is a form of recreational drug use, because tobacco contains a chemical called nicotine that can be absorbed by the lungs when a person breathes in tobacco smoke.

### Description

Smoking has been practiced by humans since at least 5000 BCE as part of offerings to divine beings or as a cleansing ritual. Most of these early forms of smoking used herbs, incense, or hallucinogens. Tobacco smoking appears to have started among the Maya and Aztecs of Mexico and Central America, first as a practice used by the priests to make contact with the spirit world, and later as a recreational activity among the Aztec nobles. By the time the Spanish conquistadors arrived in the 1520s, recreational smoking was widespread among wealthy Aztecs.

Smoking quickly spread around the world in the sixteenth and seventeenth centuries, as European traders brought tobacco from the Americas to China and Ottoman Turkey as well as to Europe itself. Nicotine, the addictive chemical in tobacco, takes its name from Jean Nicot (1530–1600), a French diplomat who introduced tobacco to the

#### Also Known As

Nicotine addiction, nicotine dependence

#### Cause

Combination of genetic and psychological factors with addictive qualities of nicotine

#### Symptoms

Inability to stop smoking; withdrawal symptoms

#### Duration

Years

#### Smoking

*Normal lung tissue (left) and one damaged by smoking.* © A. GLAUBERMAN/SCIENCE SOURCE/PHOTO RESEARCHERS, INC.



French court. One of the historical ironies of tobacco use is that smoking was originally thought to be beneficial to health. Some European doctors claimed that smoking could sober up alcoholics and cure sexually transmitted diseases like syphilis. Although Benjamin Rush (1745–1813), a signer of the Declaration of Independence, as well as a physician, argued that tobacco smoke is harmful to health as early as 1798, it was not until 1948 that a British researcher named Richard Doll (1912–2005) published proof that smoking increases the risk of lung cancer and heart disease.

In spite of antismoking campaigns since the 1970s, smoking remains a difficult habit to break. One reason is the addictive nature of nicotine itself; it is considered even more addictive than cocaine. When inhaled in tobacco smoke, nicotine reaches the brain within seconds. It stimulates the release of dopamine, a brain chemical that causes most people to feel good. Nicotine is also a stimulant—a drug that makes people feel more alert and awake. Thus people smoke in order to relieve stress, to wake up in the morning, to stay alert when tired, or as part of social get-togethers. The habits that build up around tobacco use are another reason why people find it hard to quit smoking.

Smoking is harmful to health partly because of nicotine itself and partly because of the other chemicals contained in tobacco smoke along with nicotine. Nicotine increases a person's risk of heart disease because

## The Great American Smokeout

The American Cancer Society (ACS) sponsors a program called the Great American Smokeout every year on the third Thursday of November. The aim of the Smokeout is to get smokers to stay away from tobacco for just one day in hopes that they will make a permanent commitment to stop smoking. The Smokeout started as a local activity in 1974 when a newspaper editor in Minnesota started the state's first Don't Smoke Day, or D-Day. In 1976, the California chapter of the ACS took up the idea, and in 1977 the ACS took the Great Smokeout nationwide.

The present program includes a Quitline that people can call for help in quitting twenty-four hours a day, a guide to quitting smoking successfully, a list of reasons to quit, and links to local Great American Smokeout activities. People who want help quitting can telephone 1-800-ACS-2345 to find a quitline or other science-based support group in their area. They can also ask for help via the Great American Smokeout's website at http://www.cancer.org/ docroot/subsite/greatamericans/Smokeout.asp. it raises blood pressure and speeds up the heart rate. It doubles a person's risk of stroke. Because the tobacco in a cigarette or cigar does not burn completely, the smoke contains carbon monoxide. It also contains arsenic, cyanide, and at least sixty chemicals known to cause cancer. In addition to causing 90 percent of deaths from lung cancer, smoking also causes cancers of the bladder, oral cavity, pharynx, larynx (voice box), esophagus, cervix, kidney, lung, pancreas, and stomach. Women who smoke have an increased risk of giving birth prematurely or having the baby die before birth. They are also at increased risk of osteoporosis after menopause.

## **Demographics**

Cigarette smoking is considered the leading cause of preventable illness and premature death worldwide. As of 2008, it is estimated that about 1.1 billion people around the world are smokers, more of them in the developing countries than in the West. The rates of smoking are increasing rapidly in China and India but decreasing in the United States. As of 2008, about 28 percent of American males and 24 percent of females smoke, down from 52 percent of males and 34 percent of females in 1965.

Cigarette smoking costs the United States about \$195 billion in health care costs each year, or an average of \$4,300 per adult smoker. About 438,000 Americans die each year from smoking-related diseases. More deaths are caused each year by tobacco use than by all deaths from AIDS, illegal drug use, alcoholism, motor vehicle injuries, suicides, and murders combined. In addition to the death toll, about 8.6 million people in the United States as of 2008 had at least one serious illness caused by smoking.

Risk factors for becoming a smoker include:

• Low levels of education. Forty percent of men who did not complete high school smoke, compared to 17 percent of college graduates.

- Low income.
- Race. Forty percent of African American men and 30 percent of Hispanics smoke compared to 27 percent of Caucasian men.
- Age at which a person starts smoking. People who begin to smoke before age eighteen are far more likely to be heavy smokers as adults than those who started after age twenty-one. As of 2008, the average age of first-time smokers had dropped to 14.5 years.
- Having parents who smoke. Teens with parents who smoke are twice as likely to start smoking.
- Depression. Depression is a common symptom of nicotine withdrawal. People who have become depressed may start smoking in order to soothe their feelings and then find themselves unable to quit.
- Genetic factors. Recent research indicates that some people have a gene that influences the production of an enzyme that clears nicotine from the bloodstream fairly rapidly. These people tend to smoke more heavily and find it harder to quit.

## **Causes and Symptoms**

People start smoking as a result of a combination of social and individual psychological factors. They then become dependent on smoking because of the addictive qualities of nicotine and possible genetic factors that increase their dependency on nicotine.

The symptoms of nicotine dependence include:

- Inability to quit smoking in spite of one or more serious attempts.
- Craving for a cigarette at certain times of day or in specific situations.
- Continuing to smoke in spite of lung disease, heart disease, or other smoking-related health problems.
- Withdrawal symptoms when trying to quit smoking. These may include anxiety, depression, headache, drowsiness, difficulty concentrating, diarrhea, irritability, insomnia, and weight gain.

## Diagnosis

Most visits to doctors about smoking are usually to ask for help with quitting rather than diagnosing that a person smokes, though nicotine withdrawal can be diagnosed on the basis of the patient's smoking history and the presence of withdrawal symptoms. Doctors and dentists are, however, in a good position to give their patients the facts about the harmful effects of smoking and offer encouragement and advice about quitting.

## Treatment

Treatment of smoking is called smoking cessation. The goal is to help the smoker quit smoking permanently. There are a number of different approaches to smoking cessation:

- Quitting cold turkey. This term refers to stopping without medications or other helps and relying on will power alone. About 90 percent of smokers try to stop cold turkey on their first attempt; however, only about 10 percent are able to stop for as long as six months this way.
- Counseling and psychotherapy. This approach attempts to help smokers understand their reasons for smoking and identify situations or other triggers that increase the urge to smoke. It helps about 25 percent of smokers stay smoke-free for six months.
- Antismoking medications. These include nicotine replacement gums, patches, and inhalers that work to reduce cravings for tobacco by supplying the body with nicotine in other forms. A newer drug called Chantix works by occupying the nicotine receptors in the brain even though it is not nicotine itself. Chantix reduces withdrawal symptoms and the satisfaction people get from smoking. Doctors usually suggest that smokers combine these medications with support group meetings, smoking cessation programs, or ongoing guidance from a health professional rather than relying on the drugs alone.
- Antidepressants. A drug called Zyban is sometimes prescribed to help smokers quit. It helps some people stop smoking by increasing the level of dopamine in the brain.
- Group therapy and support groups. Many smokers find support group meetings helpful in dealing with the social triggers of cigarette smoking.
- Complementary and alternative treatments. Some smokers report that hypnosis, acupuncture, and massage therapy are useful in helping to quit.

## WORDS TO KNOW

**Nicotine:** A chemical found in tobacco that acts as a stimulant in humans.

**Smoking cessation:** A term that refers to a product or program to help people quit smoking.

**Stimulant:** Any drug or chemical that temporarily increases the user's awareness or alertness.

Withdrawal: A group of physical and emotional symptoms associated with stopping tobacco use.

## Prognosis

Quitting smoking is very difficult; it is much better never to start in the first place. According to the Surgeon General, only about 30 percent of attempts to quit last longer than six months. The average person who does succeed in quitting takes between seven and fifteen attempts to do so. The benefits of quitting at any age, however, make the effort worth-while. A person who quits smoking before age thirty-five will avoid 90 percent of the health risks associated with smoking. A person who succeeds in quitting between the ages of thirty-five and fifty cuts his or her risk of dying before age sixty-five in half compared to people who continue smoking. Even someone who quits at age sixty-five will increase his or her lifespan by an average of three years.

### Prevention

Many programs to prevent smoking in the United States are aimed at teenagers, on the grounds that more than 90 percent of first-time tobacco use occurs before high school graduation, and about 40 percent of teenagers who smoke become addicted to nicotine. Among the specific measures to lower the rate of smoking among young people are:

- Imposing legal penalties for food stores or supermarkets that sell tobacco products to anyone under 18.
- Education programs in schools, patient handouts in doctors' and dentists' offices, and personal counseling by health care professionals about the dangers of smoking.
- Limiting or forbidding smoking in restaurants, offices, stores, and other public places.
- Limiting the advertising of cigarettes and other tobacco products.
- Taxing tobacco products heavily.

### The Future

As of 2008 there were several nicotine vaccines undergoing clinical trials as aids to smoking cessation. One is called NicVAX and is thought to work by stimulating the body to produce antibodies against nicotine. The antibodies block nicotine from reaching the brain, thus interfering with the pleasurable effects of smoking. Other nicotine vaccines being studied include one called TA-NIC and another called Nicotine-Qbeta. Both of these work like NicVAX in producing antibodies against nicotine.

**SEE ALSO** Alcoholism; Asthma; Bronchitis; Coronary artery disease; Depression; Emphysema; Heart attack; Heart failure; Hypertension; Laryngitis; Lung cancer; Marijuana use; Osteoporosis; Periodontal disease; Prematurity; Stroke; Tooth decay; Tuberculosis

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## **m** Sore Throat

## Definition

Sore throat, also known as pharyngitis, is a common symptom associated with upper respiratory infections, surgical procedures, and other irritants that may inflame the tissues of the throat. It is not a separate disease.

## Description

The discomfort associated with a sore throat varies somewhat according to its cause. Most sore throats are caused by viruses, specifically the viruses that cause the common cold. A person with a viral sore throat will be sneezing and having a runny nose as well as coughing and feeling a mild headache. A sore throat caused by the flu, however, will usually be accompanied by a fever of 102°F (38.8°C) or higher, and muscle aches and general tiredness. A patient with a sore throat caused by the virus responsible for infectious mononucleosis may have swollen tonsils, swollen lymph glands in the neck, and be tired for several weeks after the sore throat goes away.

A sore throat caused by bacteria, such as those that cause strep throat, may be very painful; the patient may drool or spit because swallowing is difficult. Bacterial sore throats also often come on suddenly.

Sore throats caused by smoking, air pollution, allergies, or overuse of the voice are often characterized by a dry or scratchy feeling in the throat rather than an aching sensation. Some people also experience this type of sore throat during winter, particularly in the mornings, because dry heat from a furnace tends to irritate the tissues that line the throat.

A sore throat caused by an injury to the lining of the throat—for example getting a fish bone caught in the throat or having a medical examination that requires putting an instrument down the throat usually involves pain only in the area of the injury. It is not accompanied by fever, sneezing, or other symptoms of an infection.

In rare cases, a sore throat is the first sign of throat cancer. Cancerous tumors in the throat are often accompanied by coughing up blood, pain

#### **Also Known As** Pharyngitis

#### Cause

Viruses, bacteria, allergies, tobacco smoke, air pollutants, surgery, swallowing a foreign object

#### Symptoms

Loss of voice, difficulty swallowing, fever and other symptoms of upper respiratory infections

#### Duration

A few days to two weeks, depending on the cause



A doctor uses a tongue depressor to check for a sore throat. SHUTTERSTOCK.

in the throat that does not go away with antibiotic treatment, difficulty in swallowing, and a hoarse voice.

## **Demographics**

Sore throats are one of the most common reasons for staying home from school or work. The National Institutes of Health (NIH) estimates that sore throats are responsible for forty million doctor visits every year in the United States. Children have an average of five upper respiratory infections causing sore throats every ear, and adults have between one and three. About 90 percent of sore throats are caused by one type of virus or another, with most of the remaining 10 percent being caused by bacterial infections.

Sore throats caused by viruses are equally common in both sexes and in all racial and ethnic groups in the United States.

People at increased risk of sore throats include:

- Smokers or those exposed to secondhand smoke
- People whose jobs expose them to chemicals that irritate the throat
- People with seasonal allergies to plant pollen, or year-round allergies to household dust, pet dander, or molds
- People with diabetes
- People with weakened immune systems, including those with HIV infection and those receiving chemotherapy for cancer
- People who live or work in close quarters with others, such as college students, military personnel, child care workers, office workers, and hospital staff
- People with frequent sinus infections

## **Causes and Symptoms**

Sore throat can have many possible causes, ranging from viruses and bacteria to smoking, air pollution, allergies, swallowing a foreign body, dry air, and overuse of the voice; however, most cases of sore throat are caused by viruses. The different types of viral infection that may be accompanied by a sore throat include:

- Common cold (There are about 200 different viruses that can cause colds.)
- Influenza
- Infectious mononucleosis
- Cold sores
- Measles
- Chickenpox
- HIV infection

The most common bacterial cause of sore throat is the bacterium that causes strep throat. Other bacterial infections that can cause sore throat include gonorrhea, a sexually transmitted disease, and diphtheria. Diphtheria is a potentially life-threatening infection of the upper respiratory tract that has been virtually wiped out in the developed countries since the development of a vaccine against it in the 1980s.

The symptoms that accompany sore throat depend on its cause. In general, sore throat caused by a viral or bacterial infection will look red or swollen when the doctor examines the patient's upper respiratory tract, and the tonsils will often look enlarged and have a coating of

## When to See the Doctor

Most sore throats, particularly those caused by cold viruses, go away on their own after a few days without the need for a visit to the doctor. People should, however, see their doctor if they have any of the following symptoms, which could indicate a serious condition:

- Severe difficulty in swallowing or breathing
- A sore throat that lasts longer than a week, or hoarseness that lasts longer than two weeks
- Problems sleeping because of pain in the throat
- Fever of 102°F (38.8°C) or higher
- Sunken eyes, weakness, low urine output, and other signs of dehydration
- Pus in the back of the throat
- Skin rash
- Lump in the neck
- Coughing up blood, or blood in the saliva
- Family member or acquaintance diagnosed with strep throat
- Pain in the abdomen or vomiting

pus. A sore throat caused by smoking, chemical irritants, tissue injury, or dry air will not be accompanied by swollen tonsils or pus. In the case of throat cancer, the tumor will often be visible when the doctor examines the inside of the patient's throat.

## Diagnosis

In most cases, the doctor can diagnose the cause of the patient's sore throat by taking a history (asking about recent exposure to people with colds, flu, mononucleosis, or strep throat; occupation; history of allergies; smoking habits; and similar questions), and by examining the inside of the patient's mouth and throat. It is not always easy to distinguish between a sore throat caused by a virus and strep throat just by looking, however, so the doctor will take a sample of fluid from the patient's throat on a cotton swab. The fluid can be sent to a laboratory for a throat culture, which is an accurate test that takes two days. The doctor may also perform what is called a rapid strep test in the office.

A blood test called the monospot test can be performed if the doctor thinks that the patient may have mononucleosis.

If there is a tumor in the throat, the doctor can take a sample of tissue to be analyzed for evidence of cancer. In addition, the patient will be given a computed tomography (CT) scan or magnetic resonance imaging (MRI) of the head and neck to see whether the tumor is limited to the throat or has started to spread to other parts of the body.

#### Treatment

Antibiotics are not effective in treating a sore throat caused by a virus. Most doctors prefer to wait until they have the results of a throat culture to prescribe antibiotics.

Adults or children with strep throat or a sore throat caused by colds or flu can also take ibuprofen, acetaminophen, or another nonaspirin pain reliever to bring down fever and relieve muscle cramps or headache. Gargling with salt water—a half teaspoon of salt in a glass of warm water is recommended for easing the throat discomfort.

Other remedies recommended by doctors for sore throats include:

- Using a humidifier in the home to help keep the throat tissues moist
- Drinking lots of liquids
- Drinking warm tea with honey and a small amount of lemon juice
- Getting enough sleep

The doctor may recommend a tonsillectomy (surgical removal of the tonsils) for children with recurrent severe sore throats. Tumors in the throat are treated with surgery and radiation therapy.

#### Prognosis

Sore throats caused by viral infections other than HIV usually clear up with no long-term complications once the person has recovered from the illness. Most people with strep throat also recover completely; however, about two patients per 1,000 who are not treated for strep throat will develop rheumatic fever and another two per 1,000 will develop a severe infection of the tonsils. Other possible but rare complications of strep throat include ear infections, kidney or liver damage, pneumonia, inflammation of the bones or joints, or sinusitis.

The prognosis of throat cancer depends on the stage of disease at the time of diagnosis. Early-stage throat cancers have a high cure rate.

### Prevention

It is difficult to completely prevent sore throat, particularly during cold and flu season, but people can reduce their risk by taking the following precautions:

- Wash the hands frequently, and use hand sanitizers containing alcohol when soap and water are not available.
- Do not share drinking glasses, food utensils, towels, or other personal items with others.
- Avoid close contact with people who have colds or other upper respiratory infections.
- Quit smoking (or do not start in the first place) and avoid exposure to secondhand smoke.
- Use a humidifier in the home during the winter.
- Stay indoors on high pollution or high pollen count days.
- If the sore throat is related to overusing the voice, give the voice a rest for several days.

## The Future

Because of the many different possible causes of sore throat and the near impossibility of eliminating the common cold and other viral illnesses, the condition will continue to be a frequent health problem for children and adults alike.

**SEE ALSO** Allergies; Common cold; Infectious mononucleosis; Influenza; Laryngitis; Smoking; Strep throat; Tonsillitus

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## U Spipa Difi

## Spina Bifida

## Definition

Spina bifida (pronounced SPEYE-na BIFF-ih-da) is a birth defect in which the tissues and vertebrae that normally cover the spinal cord fail to close completely during the baby's development before birth. The words *spina bifida* mean "split spine" in Latin. Spina bifida is also known as a neural tube defect (NTD). Neural tube is the medical term for the folds of tissue in a human embryo that eventually give rise to the brain and spinal cord.

#### Description

Spina bifida is a birth defect that occurs during the first few weeks of pregnancy, before the mother knows she is pregnant. As the fertilized egg develops into the embryo, it forms three layers of tissue, one of which contains cells along the back of the embryo that eventually form the central nervous system. As this group of cells grows, it folds inward to form a sheath that eventually closes to form the brain and spinal cord. This closure usually takes place around the twenty-eighth day of pregnancy. If a portion of the neural tube fails to close properly, the baby's spine may be weakened, or portions of the spinal cord and meninges (the membranes that cover the brain and spinal cord) may push through the opening in the vertebrae.

Also Known As Neural tube defect. NTD

#### Cause

Incomplete closure of the spinal column during fetal development

#### Symptoms

None (in mild cases); bowel and bladder problems, seizures, partial paralysis (severe cases)

#### Duration

Lifelong unless corrected by surgery



**Baby with spina bifida.** © BIOPHOTO ASSOCIATES/ PHOTO RESEARCHERS, INC.

The severity of spina bifida varies from a mild form that may be discovered only by accident to a severe form that can lead to premature death if untreated. There are three main types:

- Spina bifida occulta. This is the mildest form. *Occulta* means "hidden," because the condition is not noticeable at birth. In this form, there is a small space or gap in one or several of the vertebrae in the spinal column. Because there is no damage to the spinal cord or other nerves, children with spina bifida occulta do not usually have symptoms. In many cases the gap in the vertebrae is discovered only during a routine x ray study in later life. Doctors estimate that between 5 and 30 percent of cases may fall into this group.
- Meningocele. This is a relatively rare form. The meninges that cover the spinal cord may push outward through the gap in the vertebrae to form a lump or cyst. The spinal cord itself is not affected, and in most cases the cyst can be removed surgically without damage.
- Myelomeningocele. This is the most severe form, also known as open spina bifida, which usually involves some type of permanent disability. A section of the spinal cord and the nerves leading outward from it protrude through the gap in the vertebrae and are exposed on the outside of the body.

## Spina Bifida and Euthanasia

In 2004 a group of doctors in the Netherlands made headlines around the world with their publication of the Groningen Protocol, a document justifying the euthanasia, or mercy killing, of infants that the doctors regarded as involving "unbearable and hopeless suffering." The doctors then published an article in the *New England Journal of Medicine* in 2005 defending their position and discussing the euthanasia of twenty-two newborns between 1997 and 2004 on the grounds that their cases were hopeless. All had spina bifida.

The article disturbed many people because even severe spina bifida is not considered a fatal disability. None of the twenty-two infants died simply from allowing their birth defect to take its course; they were killed by administering an overdose of a sleeping medication. Another Dutch doctor openly questioned the Groningen Protocol's reference to "unbearable suffering," noting that the majority of patients with myelomeningocele can be treated surgically and have good prospects of a productive adult life. He concludes, "There is no reason whatsoever for active life-termination of these newborns." An ongoing question remains: if spina bifida is considered a reason for ending a newborn's life, might other diseases come to be regarded in the same way?

## Demographics

Spina bifida is the most common permanently disabling birth defect worldwide. It is estimated to affect one to two of every 1,000 newborns in the United States. In some parts of the world, the rates are higher: in Ireland and Wales, for example, there are three to four cases of myelomeningocele per 1,000 births.

There are some variations in frequency among different ethnic groups in the United States, with Hispanics having the highest rate of spina bifida, followed by Caucasians, Native Americans, African Americans, and Asian Americans. The total number of Americans with severe spina bifida was estimated to be around 70,000 in the early 2000s.

Spina bifida is slightly more common in girls than in boys, although the reason for this difference is not known.

## **Causes and Symptoms**

The exact cause of NTD is not known, but is thought to be the result of interactions between genetic factors and the mother's diet. The fact that the rates of NTD vary from country to country and among different races suggests that genes are involved in the disorder. In addition, researchers have found that a woman who has one child with NTD has a greater chance of having a second child with NTD. Diabetes and

seizure disorders also increase a woman's risk of having a baby with spina bifida.

With regard to diet, it is known that adequate amounts of folic acid (a form of vitamin B<sub>9</sub>) in the mother's food during pregnancy can help to prevent spina bifida, though scientists are not yet certain why the vitamin has this beneficial effect. Folic acid is found in green leafy vegetables, egg yolks, dried peas and beans, brewer's yeast, fortified cereal products, and certain other fruits and vegetables. It can also be taken by itself as a dietary

supplement. Since January 1998, following recommendations from the Food and Drug Administration (FDA), folic acid has been added to breads, breakfast cereals, and other grain products sold in the United States.

The symptoms of spina bifida vary considerably according to the severity of the defect:

- Spina bifida occulta: There are usually no symptoms of damage to the nervous system; sometimes a small birthmark or tuft of hair on the skin over the small opening in the affected vertebrae is visible.
- Meningocele. A small fluid-filled sac, usually covered by a thin layer of skin, protrudes from the baby's back.
- Myelomeningocele. There may be a sac on the baby's back at birth, but in most cases the nerves and other tissues are exposed, leaving them extremely vulnerable to infection.

Open spina bifida is associated with a number of health problems and physical disabilities, depending on the location of the exposed part of the spinal cord.

- There is usually some degree of paralysis below the level of the defect in the spinal cord. The closer the defect lies to the head and neck, the more difficulties the person will have with walking, coordination, deformities of the hips and knees, and loss of muscle tone.
- A defect close to the lower back may lead to intense pain in the lower back, upper leg, and knee.
- Defects occurring toward the base of the spinal cord often result in loss of bowel and bladder control.
- About 90 percent of patients with open spina bifida develop hydrocephalus, or a buildup of fluid inside the skull caused by interference with the normal flow of cerebrospinal fluid, potentially lead to learning difficulties and problems with concentration as the child grows older.
- Babies with open spina bifida are at increased risk of meningitis, an infection of the meninges that can be life-threatening.
- Tethering of the spinal cord. With this complication, the spinal cord cannot move freely inside the vertebrae but is attached (tethered) to nearby tissues, which can lead to scoliosis (curvature of the spine), dislocation of the hip, and other deformities.
- Latex allergies, depression, obesity, and stomach disorders. These complications occur as children with spina bifida grow older.

### Diagnosis

Spina bifida can be diagnosed during pregnancy by testing the mother's blood for a protein called alpha-fetoprotein, which is produced by the fetus and the placenta during pregnancy and enters the mother's blood-stream in small amounts. Unusually high levels of this protein may indicate a neural tube defect. The doctor can order additional tests, including taking a sample of the amniotic fluid surrounding the baby and performing an ultrasound imaging study.

Mild cases of spina bifida can be detected after the baby is born by an x-ray study, computed tomography (CT) scan, or magnetic resonance imaging (MRI). A CT scan may be ordered to check for fluid inside the skull if the doctor thinks that the baby may have hydrocephalus.

## Treatment

There is no cure for spina bifida. Treatment depends on the location and severity of the defect in the spine. Babies with spina bifida occulta may not need any treatment, and those with meningocele are usually treated by having the cyst on the back removed by surgery. They usually have no complications.

Treatment of myelomeningocele is complex, as children with this severe form of spina bifida usually need extensive physical therapy and sometimes special education following surgery. In most cases newborns with open spina bifida are operated on as soon as possible after birth. They are given antibiotics to prevent infection of the exposed part of the spinal cord. The opening of the spinal cord is closed, and covered with skin and muscles taken from either side of the back. If the baby has hydrocephalus, a shunt system may be inserted to redirect excess fluid away from the brain to other parts of the body.

As the child grows older, additional operations are usually needed to correct a tethered spinal cord or scoliosis or to replace the shunt for hydrocephalus. Physical therapy usually includes range-of-motion exercises as well as strength exercises to prepare the child for walking with braces or crutches.

## Prognosis

The prognosis for patients with spina bifida occulta and meningocele is good. The prognosis for patients with myelomeningocele depends on the location of the defect in the spinal column and the amount of spinal cord

## WORDS TO KNOW

- **Euthanasia:** Sometimes called mercy killing; the act of killing a hopelessly ill human or pet in a painless way.
- **Folic acid:** A form of vitamin B<sub>9</sub> that helps to prevent spina bifida.
- **Meninges (singular, meninx):** The three layers of protective tissue that cover the central nervous system.
- **Neural tube:** The medical term for the folds of tissue in the human embryo that eventually form the brain and spinal cord.
- **Shunt:** A flexible plastic tube inserted by a surgeon to drain cerebrospinal fluid from the brain and redirect it to another part of the body.

that was exposed. Advances in prenatal diagnosis and surgery mean that most patients survive; and early treatment of hydrocephalus prevents mental retardation in most cases. With appropriate follow-up surgery and physical therapy, most patients with open spina bifida can complete their education and live independently; many marry and have families of their own.

## Prevention

Although the genetic factors that may be involved in spina bifida have not been identified, adding folic acid to a mother's diet has been found to reduce a the baby's risk of spina bifida by 70 percent.

## The Future

New approaches to surgical treatment of spina bifida include fetal surgery or operating on the baby before delivery. This technique involves opening the mother's abdomen and uterus and closing the opening over the baby's spinal cord. Fetal surgery is usually performed between the nineteenth and twenty-fifth weeks of pregnancy. Some researchers think that this early surgery improves the baby's chances of normal brain development as well as lowering the risk that the spinal cord may become infected; however, it also increases the risk of premature birth and the baby's death.

Other areas of research include the search for specific genes associated with spina bifida and studies that may shed light on why folic acid helps to prevent it.

SEE ALSO Hydrocephalus; Meningitis; Scoliosis

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## Spinal Cord Injury

## Definition

Spinal cord injury, or SCI, refers to damage caused either directly to the cord itself or indirectly by injury to the bones, muscles, or soft tissues surrounding the cord. Most spinal cord injuries are caused by accidents or other traumas, but some are the result of tumors, birth defects, or diseases that affect the spine or surrounding muscles. The spinal cord can be compressed by damage to the vertebrae surrounding it or by blood and tissue fluid accumulating inside the spinal column. The spinal cord can also be completely cut or torn in an accident or by a knife or gunshot wound.

#### Also Known As

Spinal cord trauma, SCI

#### Cause

Direct or indirect injury to the spinal cord

#### Symptoms

Weakness, numbness, pain, paralysis, loss of bowel and bladder control

#### Duration

Years

Spinal cord injuries can be classified in two ways, by degree of severity and by location. Some doctors refer to an SCI as either complete, meaning that the patient has no sensation or ability to move muscles below the injury; or incomplete, meaning that the patient has some degree of sensation or function in the parts of the body below the injury.

The location of a spinal cord injury is identified by the number of the affected vertebra. The human spine is divided into four segments: cervical (the neck region, containing seven vertebrae); thoracic (the chest region, containing twelve vertebrae); lumbar (the lower back, containing five vertebrae); and sacral (the tailbone region, containing five vertebrae). Thus a C4 injury refers to the fourth cervical vertebra; T12 refers to an injury at the level of the twelfth thoracic vertebra; and so on.



## Description

Injury to the spinal cord at any point along its length involves a series of events that damage the nerve endings in the cord. The spinal cord contains two major types of neurons, or nerve cells: sensory neurons that carry messages from the skin, joints, and muscles upward to the brain; and motor neurons, which carry messages from the brain downward to the various parts of the body. Thus any injury to the cord interferes with both sensation and movement.

When the spinal cord is compressed, torn, or cut, the cord swells to fill the entire inside of the spinal column, which cuts off the blood supply to the injured tissue. The patient's blood pressure drops and the nerve endings lose their ability to transmit electrical impulses to or from the brain. This condition is called spinal shock. It is followed by a secondary phase of damage that includes inflammation, the overstimulation of injured nerve cells, and the self-destruction of these nerve cells.

## Demographics

According to the National Institutes of Health, there are between 10,000 and 12,000 spinal cord injuries each year in the United States. As of

Image of a spinal cord injury in a human neck. The spinal cord (pink) is pinched by the back bones at the center of the picture. LUNAGRAFIX / PHOTO RESEARCHERS, INC.

## First Aid for Spinal Cord Injuries

A spinal cord injury is a medical emergency. It is important, however, *not to move* someone who may have an SCI as paralysis or permanent injury could result. A spinal cord injury should be assumed in any of the following circumstances

- The person has a head injury and is drifting in and out of consciousness.
- The injured person complains of pain in the neck or back.
- The person cannot move their neck.
- The injury to the head or neck was caused by significant force.
- The person's neck or back is twisted or is positioned oddly.
- The person complains of weakness, numbness, or paralysis.
- The person cannot control their limbs, bladder, or bowels.

To help someone with a suspected SCI:

- Call 911 at once.
- Keep the person still. Put heavy towels on both sides of the neck or hold the head and neck to prevent movement. It is important to keep the person in the same position as they were in when found until emergency help arrives.
- Give as much first aid as possible without moving the injured person's head or neck.
- If it is absolutely necessary to turn the injured person to one side because they are choking on blood or vomiting, get someone else to help turn them. Work together to keep the injured person's head, neck and back in alignment while rolling them onto one side.

2008, there were 255,000 Americans living with these injuries; their care costs the country about four billion dollars per year.

Car accidents are the largest single cause of spinal cord injuries, being responsible for 42 percent. Another 27 percent result from falls; the remainder are caused by athletic accidents (8 percent), criminal violence (15 percent), and work-related injuries (7 percent).

There are four times as many men with spinal cord injuries as women. Fifty-five percent of these men are between sixteen and thirty years old.

## **Causes and Symptoms**

Spinal cord injuries may be caused by transportation accidents, falls, workplace accidents, violence, or diseases that affect the bones or muscles of the spinal column.

The symptoms of spinal cord injury include:

- Pain or intense stinging sensations at the point of injury
- Increased muscle tone or muscle spasms
- Inability to feel heat, cold, or touch
- Loss of bowel or bladder control
- Loss of the ability to move parts of the body below the point of injury
- Difficulty breathing or coughing
- Problems with blood pressure or temperature regulation
- Abnormal sweating

The specific disabilities that may result from spinal cord injury depend on the location of the damage. In general, the higher the level of injury (closer to the head), the more extensive the paralysis. Some patterns of disability are as follows:

• C1 through C3: Injuries to these neck vertebrae result in quadriplegia, or paralysis of both arms and both legs. The patient will need to be placed on a respirator in order to breathe.

- C5 and C6: The person has some arm function but not the use of the hands.
- T1 through T8: The person has paraplegia; that is, he or she cannot use the legs or lower part of the trunk. The hands, arms, head, and breathing are usually not affected.
- T9 through T12: The person can control their abdominal muscles and sit upright.
- L1 through L5: The person will have difficulty bending or flexing the legs and hips.
- S1 through S5: The person may have some difficulty with bowel and bladder control.

## Diagnosis

A person with a suspected SCI must be moved carefully by a specially trained trauma team. The usual practice is to fit a stiff collar around the injured person's neck and move him or her on a rigid board, to prevent further injury to the spinal cord. The injured person is taken to the hospital as quickly as possible, as it is critical not to delay treatment.

If the injured person is able to talk, the doctor may be able to determine the location of the injury fairly rapidly by asking about pain and other sensations, lack of ability to move parts of the body, and so on. The doctor may also test the patient's reflexes or use pinpricks or other forms of touch to determine whether sensation has been lost. If the patient is having difficulty breathing or has lost consciousness, the doctor will order one or more imaging studies, including x rays, computed tomography (CT) scans, and magnetic resonance imaging (MRI). Another imaging technique that is often used with spinal cord injuries is myelography. This test consists of injecting a contrast dye into the spinal column that will show up on an x ray or CT scans.

Patients are usually given a second round of diagnostic tests and imaging studies a few days after the injury to evaluate the extent of the injury and the patient's chances for recovering sensation and function.

## Treatment

Treatment of spinal cord injuries usually involves a combination of medications, surgery, traction or bracing, and long-term physical therapy and rehabilitation.

- A medication called Medrol can be given to patients with a severe SCI. Medrol is a corticosteroid that reduces swelling and inflammation of the spinal cord. It must, however, be given within eight hours of the injury.
- Surgery is performed to remove bone fragments, foreign objects, or vertebrae that are compressing the spinal cord. Surgeons differ as to whether it is better to perform surgery immediately or to wait until the extent of the patient's injuries has been determined.
- Traction and braces or a body harness are applied to bring the patient's spine into proper alignment during healing.
- Physical therapy and rehabilitation. These forms of therapy usually begin after the patient has left the hospital, which may take from a few days to several weeks. Rehabilitation includes learning to cope with bowel, bladder, or sexual functions (if any), self-care, and the use of a wheelchair or other assistive devices as well as regaining muscle strength and range of motion.

Most patients also need psychotherapy and support groups to cope with depression and other emotional problems that often occur after a spinal cord injury, particularly if the patient is quadriplegic or paraplegic.

## Prognosis

The prognosis of a spinal cord injury varies. It may take several weeks or months for the patient's doctors to determine the extent of possible recovery. In general, impairment that remains a year after the injury is likely to be permanent. Some studies indicate that age makes a difference in a patient's ability to regain function; younger patients are more likely to improve than patients over fifty years of age.

A common complication of spinal cord injuries is chronic pain. Two-thirds of patients with SCIs have ongoing pain, with half of these reporting that their pain is severe. There is no universally successful treatment for such pain. Most patients are given medications for the pain, usually antidepressants or antiseizure drugs.

Spinal cord injury shortens most patients' life spans, although people still live longer with such injuries than was the case in the 1960s. Pneumonia is a common cause of death in patients with quadriplegia. Another common cause of death is suicide; patients with spinal cord injuries have a suicide rate five times higher than the general population.

## WORDS TO KNOW

**Paraplegia:** Paralysis that affects only the lower body.

**Traction:** The use of braces, casts, or other devices to straighten broken bones and keep them aligned during the healing process.

**Quadriplegia:** Paralysis that affects both arms and both legs. It is also known as tetraplegia.

## Prevention

Many spinal cord injuries can be prevented by taking basic safety precautions:

- Practice safe driving. This includes the use of seat belts, keeping the car in good repair, and avoiding driving after drinking or taking drugs.
- When swimming, check the depth of water before diving. Be particularly careful when diving in natural bodies of water because of the possibility of hidden rocks.
- Use appropriate protective equipment when playing football or other contact sports. Avoid movements that put the head and neck at risk, such as sliding into a base headfirst.
- Store firearms in a locked cabinet or safe, and store ammunition in a separate location.
- Protect against falls by checking the house or apartment for safety hazards, wearing properly fitted shoes, and reducing the use of medications that cause drowsiness or loss of balance.

## **The Future**

As of 2008, Medrol was the only drug that is generally used to bring down inflammation following an SCI, although another medication called GM-1 ganglioside is being tested as a treatment for the damage to nerve tissue that follows spinal shock. Other areas of research include various ways to stimulate the regrowth of damaged nerve fibers; the use of computers combined with electrodes implanted in the injured person to restore function to paralyzed limbs; and better ways to control pain in survivors of SCIs.

**SEE ALSO** Fractures; Multiple sclerosis; Osteoporosis; Pneumonia; Spina bifida

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## Sprains and Strains

## Definition

Sprains and strains are commonplace injuries of the joints and muscles. A sprain is a stretching or tearing of a ligament—one of the tough bands of fibrous tissue that connects bones to other bones. A strain is an injury to a muscle or tendon. Both types of injury can occur in various parts of the body, depending on the location of an injury or the part of the body that has been overused.

The most common locations for sprains are the ankle, knee, or thumb. The most common cause of an ankle sprain is called an inversion injury; it occurs when the foot turns inward when a person falls

Also Known As Pulled muscles. twisted ankle

#### Cause

Direct or indirect blow to affected area; fall; muscle overuse; inadequate rest

#### Symptoms

Pain, swelling, bruising, difficulty using joint, muscle weakness or cramping

### Duration

Days to months





A patient with a bruised and swollen sprained ankle. © PHOTOTAKE INC. / ALAMY.

on the ankle or lands on it after a jump. Less commonly, the ankle is injured when the foot turns outward suddenly; this is called an eversion injury. Knee sprains are most likely to occur as the result of a blow to the knee or falling on the knee. Thumb sprains are most likely to occur in skiers.

Strains most commonly affect the muscles of the back; the hamstring muscle at the back of the thigh; the muscles of the hand and forearm; and the muscles of the upper arm and elbow.

Sprains can be classified according to severity as well as location. Doctors usually distinguish three levels of sprains:

- Mild (sometimes called first-degree sprain): The ligament is mildly stretched or has a minor tear.
- Moderate (second-degree sprain): The ligament is torn but not completely ruptured.
- Severe (third-degree sprain): One or more ligaments are completely ruptured.

Strains can be categorized according to the type of damage caused to the injured muscle:

- The muscle tissue may tear.
- The area where the muscle joins its tendon may tear.
- The tendon itself may tear or rupture.

## When to Call the Doctor for a Sprain

Mild sprains and muscle strains can be treated at home using the PRICE therapy described in the body of the article. More severe injuries, however, require a doctor's diagnosis and treatment. A person with any of the following symptoms should see a doctor or go to the emergency room quickly:

- The person is in severe pain.
- The person cannot put any weight on the affected joint, or walk more than four steps without severe pain.
- The joint cannot be moved.
- The limb buckles or collapses when the person tries to use the affected joint.
- There is numbness in any part of the injured area.
- The affected joint or muscle has been injured several times before.
- There is redness spreading outward from the injury.
- The person has any doubt about the severity of the injury or how to care for it.

## Description

The experience of a sprain may vary from mild discomfort lasting a few minutes or hours to inability to walk or use the affected joint. In many cases the person will feel a popping or tearing sensation in the joint:

- First-degree sprain: The affected joint feels sore and hurts when it is moved, but there is not much swelling and the person can put weight on the joint.
- Second-degree sprain: The affected joint is sore, swollen, and difficult to move. There may be bruising from blood leaking into the joint. The person may feel shaky or unsteady if they try to put weight on the joint.
- Third-degree sprain: The affected joint is very painful, bruised, and swollen. The person may not be able to move it at all, and the injury may be difficult to distinguish from a bone fracture or dislocation.

A strain is usually experienced as pain in the area of the injured muscle along with cramping or spasms in the muscle, limited range of motion, and weakness in the muscle. There may also be swelling and inflammation in the affected area.

## Demographics

Sprains and strains are very common injuries in the general population; according to the National Institutes of Health (NIH), there are about 25,000 ankle sprains each day in the United States. Although amateur and professional athletes are somewhat more likely to be injured, anyone whose occupation involves lifting heavy objects, repetitive movements, or the use of power tools may develop sprains or strains. Elderly people and others at risk of falls are also more likely to sprain the knee or ankle joints. Sprains and strains are equally common in both sexes and in all races and ethnic groups.

Some people are at increased risk of sprains and strains:

- People who are in poor physical condition or who exercise when they are tired or unwell.
- People who are obese.
- People who are not using the proper techniques for their sport or occupation. Dancers, skaters, or skiers who have not been taught to jump and land properly, for example, are more likely to sprain an ankle or knee.
- People who do not warm up before athletic activities.
- People with diabetes.
- People who take medications that make them drowsy or affect their sense of balance.
- People with a history of repeated joint or muscle injuries.

### **Causes and Symptoms**

The causes of sprains and strains include accidental falls, sports injuries, overuse of weak or injured muscles, improper techniques for lifting or carrying heavy objects, repetitive use of muscles without adequate rest, or unusual stresses on a normally healthy joint or muscle.

The symptoms of sprains and strains have already been described.

### Diagnosis

Mild sprains and strains can usually be diagnosed by a primary care doctor by taking the patient's history and examining the affected joint and the tissues surrounding it. The doctor may refer the patient to an orthopaedics specialist for further evaluation of the injury. X rays may be taken to rule out broken bones but are not usually helpful in diagnosing soft tissue injuries. The doctor may order a magnetic resonance imaging (MRI) test for better evaluation of injuries to ligaments and muscles.

Numbness in the affected joint may indicate nerve damage, while coldness may indicate a problem with circulation. Either of these symptoms, or the possibility that a bone is broken or dislocated, means that the patient should go to an emergency room as quickly as possible.

## Treatment

Mild sprains and strains can be treated at home by using the PRICE approach for twenty-four to forty-eight hours:

- Protection. Protecting the injured joint or muscle involves avoiding unnecessary movement of the affected area.
- Rest. Rest means avoiding activities or body movements that make the pain or swelling in the joint worse; it doesn't mean complete bed rest.
- Ice. An ice pack can be applied to the affected area for fifteen to twenty minutes every four to six hours for one to two days.
- Compression. Compression refers to the use of a wrap, air cast, or Ace bandage to keep swollen muscles from restricting movement in an injured joint.
- Elevation. Raising the affected arm, leg, elbow, or wrist on a pillow or cushion (above heart level) is helpful in relieving swelling in the affected muscle or joint.

Nonsteroidal anti-inflammatory drugs, or NSAIDs, can be used to relieve pain and inflammation in the sore joint or muscle. These drugs include aspirin as well as Tylenol and Advil.

Moderate and severe injuries require a combination of immobilizing the affected joint with a splint or cast followed by physical therapy and rehabilitation to restore muscle strength and range of motion in the affected joint. In some cases, surgery may be needed to repair a ruptured ligament or muscle.

## Prognosis

The prognosis for recovery from sprains and strains is very good with appropriate medical treatment and self-care at home. It is important, however, for patients to follow the doctor or physical therapist's recommendations carefully to avoid reinjury. A mild ankle sprain may require several weeks of rehabilitation; a moderate sprain could require two to three months. With a severe sprain, it can take as long as eight to twelve months to return to full activity. These time frames may be even longer in elderly patients.

## Prevention

People can reduce their risk of sprains and strains by taking the following precautions:

• Avoiding sports or physical exercise when tired, ill, or in physical pain.

## WORDS TO KNOW

**Eversion injury:** An ankle injury caused when the foot is suddenly forced to roll outward.

**Inversion injury:** A type of ankle injury caused when the foot is suddenly forced to roll inward.

**Ligament:** A tough fibrous band of tissue that joins bones together.

# **Orthopaedics (also spelled orthopedics):** The branch of medicine that diagnoses and treats disorders of or injuries to the bones, muscles, and joints.

- **Tendon:** A thick band or cord of dense white connective tissue that attaches a muscle to a bone.
- Maintaining a healthy weight and eating a nourishing diet that keeps muscles strong.
- Wearing shoes that fit properly, and replacing athletic shoes as soon as the tread is worn or the heel wears unevenly.
- Doing daily stretching exercises. Yoga is a good low-impact way to stretch muscles.
- Warming up and stretching before participating in vigorous sports.
- Wearing appropriate protective equipment for baseball, football, hockey, and other similar sports.
- Being in good physical condition before playing sports.
- If running, running on even rather than rough or irregular surfaces.
- Putting safety measures in place to prevent falls, such as keeping stairways, walkways, yards, and driveways free of clutter and well lighted; making sure that rugs are anchored and electrical wires are secured; putting grab bars in shower stalls; and salting or sanding icy sidewalks and driveways in the winter.

## The Future

Sprains and strains are likely to continue to be commonplace health problems given that so many sports and occupations put people at risk for injuries to muscles and joints.

SEE ALSO Fractures; Obesity; Tendinitis

## For more information

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# Staph Infection

#### Definition

*Staphylococcus* is the name of a group of bacteria responsible for a number of serious illnesses, although most species are harmless to humans. Staphylococci are widespread in all parts of the world. They are commonly found in the soil as well as on the bodies of humans and domestic animals. Staphylococci can live on or in humans without necessarily causing harm. They can, however, cause disease in humans and other animals either by direct destruction of tissue or by releasing toxins into the digestive tract or bloodstream. Under a microscope, these bacteria look like clumps or clusters of grapes.

## Description

There are four species of staphylococci that cause various types of infections in humans. The two most important of these are *Staphylococcus* 

#### Also Known As

*S. aureus* infection, MRSA infection, staphylococcal infection

#### Cause

Bacteria belonging to the genus *Staphylococcus* 

#### Symptoms

Depends on the part of the body infected

#### Duration

Days to months, depending on location and severity of the infection *aureus*, a golden-yellow bacterium, and *Staphylococcus epidermidis*, a species that causes skin infections. *S. aureus* is generally considered the most dangerous species of staphylococcus, causing a range of infections from pneumonia and endocarditis to food poisoning and eye infections. Methicillin-resistant *S. aureus*, or MRSA, has become a major public health concern since the 1990s (see sidebar).

The other two types of staph that cause disease in humans are responsible for urinary tract infections in sexually active women and infections of the bones and joints.

## Demographics

Staphylococci are commonplace organisms found on the scalp, skin (particularly the armpits and genital areas), or outer nasal passages of humans. Biologists refer to the formation of groups or clumps of bacteria on a human or animal as colonization. Staph is found in 80 percent of the general population from time to time and 20–30 percent of the population on an ongoing basis. People who harbor staphylococci

most of the time are called "staph carriers." An estimated 2 billion people are colonized by some form of *S. aureus*; of these persons, as many as 53 million, or 2.7 percent of carriers, are thought to carry MRSA, the drug-resistant form of *S. aureus*. It is possible for a person to carry staphylococci for many years without becoming sick. In addition, such domestic animals as cats, dogs, chickens, and horses can carry MRSA strains as well as less powerful staphylococci.

As far as is known, people of either gender, any age group, or any race are equally likely to carry staphylococci. Newborns may be colonized by staphylococci from the mother during childbirth. Some groups, however, are more susceptible than others to staph infections, including diabetics, African Americans, gay men who practice anal intercourse, very young children, elderly adults, and persons with artificial joints or heart valves.



Methicillin resistant Staphylococcus aureus (MRSA) infection. © SCOTT CAMAZINE / ALAMY.

## Methicillin-resistant Staphylococcus aureus (MRSA)

Methicillin-resistant *Staphylococcus aureus* (MRSA) is a strain of the bacterium responsible for severe and potentially fatal skin and soft-tissue infections. There are two major subgroups of MRSA, named for the locations where people can get infected: community-acquired MRSA (CA-MRSA) and hospital-acquired (or healthcareacquired) MRSA (HA-MRSA).

MRSA was first identified as a particular strain of *S. aureus* in 1961. Methicillin, an antibiotic similar to penicillin, was introduced in 1959 to treat penicillin-resistant strains of *S. aureus*, but only two years later, the first strains of MRSA were reported in the United Kingdom. MRSA infections were uncommon until the 1990s, however, when their rate shot upward, particularly in hospitals.

CA-MRSA causes a boil or skin infection in about 75 percent of cases and is easily mistaken for a

spider bite. The affected area is red, swollen, and may ooze pus. CA-MRSA can be much more powerful than hospital-acquired MRSA, however, and can lead to sepsis (generalized infection of the entire body), bacteremia (infection of the bloodstream), or pneumonia. HA-MRSA is most commonly found in patients in healthcare settings, particularly those in dialysis centers, nursing homes, or other hospital settings. Patients with HA-MRSA are more likely to develop pneumonia, infected joints, or urinary tract infections than skin infections.

MRSA infections cannot be treated with the antibiotics used for most staph infections. Newer drugs like vancomycin, linezolid, or tigecycline must be used instead. As of 2008 some 94,000 serious MRSA infections were reported in the United States annually and 19,000 deaths—more than are caused by AIDS.

#### **Causes and Symptoms**

The causes and symptoms of staph infections vary somewhat depending on the species of staph involved and the specific tissues or organs affected:

- Skin and soft tissue infections. These may be caused by either *S. aureus* or *S. epidermidis*. Skin infections caused by staph often look like spider or other insect bites. Infected wounds or surgical incisions typically ooze pus or another discharge. Many of these skin infections develop from scratching insect bites or patches of eczema. After the skin is broken, the bacteria can enter the tissues beneath the surface and form large pus-filled abscesses. In patients with weakened immune systems, the staphylococci can then enter the bloodstream.
- Endocarditis. Endocarditis is an inflammation of the valves and other tissues lining the heart, caused when staphylococci form colonies on the surface of the valves. It is more common in patients

who have damaged or artificial heart valves. A patient with staphylococcal endocarditis will run a fever, and will have a heart murmur or some other abnormality of blood flow in the heart that can be detected on an echocardiogram.

- Pneumonia. Most cases of bacterial pneumonia are caused by streptococci, but about 3 percent of cases of pneumonia acquired outside hospitals are caused by *S. aureus*. These patients have the typical symptoms of pneumonia: fever, chest pain, cough, and production of sputum (mucus or phlegm from the lungs).
- Joint infections. Staphylococcal infections of the joints are common complications of patients who have artificial joints. *S. aureus* and *S. epidermidis* are the species usually involved in joint infections. The most common symptom of a joint infection is sudden swelling and pain due to pus and tissue fluid building up in the affected joint. The patient may or may not have a fever.
- Eye infections. People can get staph infections of the tissues on the inside of the eye following eye surgery or an injury to the eye. This type of eye infection is called endophthalmitis. The early symptoms of staphylococcal endophthalmitis include pain, swelling, and redness in the affected eye and partial loss of vision within a week of the injury or operation. The patient may also have a headache and be sensitive to bright light.
- Food poisoning. Staphylococcal food poisoning is the result of toxins secreted by the organisms rather than by tissue damage caused by the bacteria themselves. The foods most likely to be contaminated are those made by hand, those that require little or no cooking—such as sandwiches, cold cuts, and certain types of pastry—and those that have not been refrigerated. The symptoms of staphylococcal food poisoning include nausea, vomiting, and diarrhea, beginning between one and six hours after eating the contaminated food. Most people with staphylococcal food poisoning feel better in one to two days.

## Diagnosis

The diagnosis of a staphylococcal infection is based on a combination of the patient's medical history, symptoms, an examination of the skin or other affected body parts, and a blood culture that is positive for a specific staphylococcus species. Samples for a staph culture may be obtained from a skin injury, from drawing a blood sample, from a urine sample, or by having the patient cough up sputum if pneumonia is suspected. Although a standard blood culture for a staph infection takes a day or two to yield results, rapid diagnostic methods using amplification and probe-based molecular techniques provide results in hours, thus allowing treatment to be started earlier and improving the patient's chances of recovery. Although staph can be identified in stool samples or vomit from a patient with food poisoning, doctors do not usually test for the organism unless there is an outbreak involving several people. The diagnosis of staphylococcal food poisoning is usually made on the basis of the patient's symptoms.

## Treatment

Treatment of a staph infection depends on its specific type and location. In most cases the doctor will start antibiotic therapy when a staphylococcal infection is suspected as soon as the sample of tissue, blood, sputum, or urine has been sent to the laboratory. Specific types of infections are treated as follows:

- Skin infections: The doctor may make an incision to drain the pus and other infected fluid out of the wound. In some cases an antibiotic cream or lotion may be applied after the wound has been cleansed, or the patient may be given oral or intravenous antibiotics.
- Staphylococcal pneumonia following influenza is usually treated with intravenous antibiotics following hospitalization. People who are seriously ill may need to be given supplemental oxygen in an intensive care unit (ICU).
- If the infection is located in a joint with a prosthetic appliance, the artificial joint must be removed and the patient given a four- to six-week course of antibiotics. Infected joints without a prosthetic appliance are usually drained of fluid and the patient is given a four-week course of antibiotic therapy to clear the infection.
- Infected artificial heart valves may or may not require removal. Endocarditis does, however, require long-term antibiotic therapy, particularly if the patient is over fifty-five.
- Staphylococcal eye infections require emergency treatment. An ophthalmologist (specialist in eye disorders) usually injects antibiotics

into the tissues around the eye and also gives antibiotics by mouth or intravenously. In extreme cases the entire eye may need to be removed.

Staphylococcal food poisoning is treated with bed rest, plenty of fluids, and anti-nausea drugs prescribed by the doctor. Antibiotics cannot be used to treat food poisoning caused by staphylococci because the toxins that cause the nausea and vomiting are not affected by these drugs. Severely ill patients may need to be hospitalized and given intravenous fluids.

## Prognosis

The prognosis of staphylococcal infections varies according to the specific illness. Untreated *S. aureus* infections of the bloodstream can have a mortality rate as high as 80 percent. Endocarditis and pneumonia caused by antibiotic-resistant staphylococci have mortality rates around 11 percent in patients without other diseases or disorders, but the rate may be as high as 44 percent in patients with diabetes, HIV infection, or other disorders that weaken the immune system. Elderly people with staphylococcal pneumonia have a worse prognosis than younger adults. In patients over the age of seventy, community-acquired staph infections are associated with a mortality rate of 21 percent in the year following diagnosis.

Most patients with staphylococcal food poisoning or staphylococcal urinary tract infections recover completely; fatalities are rare except in the elderly or people with AIDS.

## Prevention

People can help to prevent staphylococcal infections by taking the following precautions:

- Avoid scratching insect bites or other areas of irritated skin; see a doctor about a boil filled with pus or a similar skin lesion that will not heal.
- Wash hands carefully before and after preparing food.
- Cover infected skin or skin draining pus with waterproof dressings, and dispose of soiled dressings carefully. Clean cuts and scratches promptly and keep them bandaged.
- People with infections on their hands or wrists should avoid preparing or serving food until the infection has been cleared.

#### Staph Infection

## WORDS TO KNOW

- **Bacteremia:** The presence of bacteria in the bloodstream.
- **Colonization:** The process by which bacteria form colonies in or on the bodies of humans and other animals.
- **Endocarditis:** An inflammation of the tissues lining the inside of the heart and its valves.
- **Endophthalmitis:** Inflammation of the tissues inside the eyeball.

- Sepsis: The presence of bacteria or their toxic products in the bloodstream or other tissues, leading to inflammation of the entire body.
- **Sputum:** Matter from the lungs or throat that is brought up by coughing.
- Strain: A genetic variant or subtype of a bacterium.
- Avoid sharing such personal items as combs, brushes, cosmetics, cell phones, razors, and towels. Be particularly careful in gyms and health clubs, as staphylococci prefer warm, moist environments.
- Wipe down kitchen countertops, athletic equipment, and hospital equipment with alcohol-based sanitizers.
- Avoid direct contact with other people's wounds or injuries whenever possible.

#### The Future

Staph infections are likely to be common for the foreseeable future because these bacteria are widespread in all countries. Research at present is focused on finding new drugs that will be effective against MRSA and improving techniques to prevent its spread outside as well as inside hospitals.

**SEE ALSO** Pneumonia; Toxic shock syndrome

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# m

# Steroid Use

## Definition

Steroid use usually refers to the abuse of synthetic hormones to build muscles, raise athletic performance, or improve one's appearance. There are, however, two major types of steroids that are used in medicine. The synthetic hormones used to build muscles are called anabolic or androgenic steroids. They are human-made drugs related to male sex hormones or androgens.

The other group of steroid medications are known as corticosteroids or glucocorticoids. They are synthetic forms of hormones that are produced naturally by the adrenal glands when the body is stressed. Corticosteroids are often prescribed to treat inflammation in disorders like eczema, arthritis, or asthma, or to treat immune system disorders like lupus or multiple sclerosis. Although corticosteroids are strong drugs with side effects and require a doctor's prescription, they cannot be used for muscle building. The remainder of this entry will focus on anabolic steroids.

## Description

Anabolic steroids are synthetic drugs modeled on the chemical structure of male sex hormones. Anabolic steroids boost the rate of protein Also Known As Steroid abuse

#### Cause

Abuse of anabolic steroids to improve one's looks or athletic performance

#### Symptoms

Voice deepening, body hair growth (women); testicle shrinking (men); stunted growth; aggressiveness

#### Duration

As long as the user continues using, typically months to years

Robert Hazelton, who claims he lost his legs due to steroid use. AP IMAGES.

formation in cells, which is why they are effective in building muscle tissue and increasing physical strength. These compounds were first made in European laboratories in the 1930s; they were recommended in bodybuilding magazines as early as 1938. Anabolic steroids were given to German soldiers during World War II (1939–1945) to increase their aggressiveness, and Adolf Hitler (1889–1945) asked his personal physician for steroid injections.

Anabolic steroids do have legitimate uses in treating bone diseases in older men as well as the wasting of muscle tissue that occurs in AIDS and certain types of cancer. They are also given to boys with delayed puberty to help them mature. What is controversial about these drugs is their use (and abuse) in amateur and professional sports to improve performance. Anabolic steroids have serious psychological as well as physical side effects in females as well as males, and can lead to long-term damage to the heart and liver as well as unattractive changes in appearance.

Anabolic steroids can be taken by mouth in the form of pills or by injections. More recently, some users have tried using steroids in the form of transdermal patches, which deliver the drug into the body through the skin.

People who use anabolic steroids have several methods for trying to speed up the effects of the drugs on building muscle. One method, called stacking, involves using several different steroids at the same time. The other method, called cycling, involves taking the drugs for six or twelve weeks at a time and not using the drugs for brief periods in between cycles. Neither stacking nor cycling makes steroids more effective, however.

#### Demographics

One reason for concern in the early 2000s is the young age of steroid abusers, as well as their numbers. Although the average age of people who use anabolic steroids for nonmedical purposes is twenty-five, about 2.7 percent of high school students are reported to use these drugs. Boys are more likely than girls to use steroids, and athletes are more likely to use them than high schoolers who are not athletes. According to a study conducted by the National Institutes of Health (NIH) in 2002, 2.5 percent of eighth graders have tried steroids; 3.5 percent of tenth graders; and 4 percent of high school seniors. College students are less likely than high schoolers to use steroids—about 1 percent.

Males between the ages of nineteen and forty are the heaviest users of anabolic steroids in the United States as of the early 2000s. The American College of Sports Medicine (ACSM) conducted a survey in 2005 of 500 young adults who use anabolic steroids. The survey found that most steroid users (78 percent) are noncompetitive bodybuilders and nonathletes who use the drugs to improve their appearance rather than to gain an edge in sports competitions. Thirteen percent of these users reported such unsafe injection practices as reusing needles, sharing needles, and sharing multidose vials of injectable steroids.

#### **Causes and Symptoms**

The cause of nonmedical steroid use in adolescents and young adults is usually dissatisfaction with one's appearance or a strong desire to excel at sports. One former user interviewed by an NIH researcher said, "When I walked into a room, I wanted heads to turn.... My whole priority was, I wanted people to say, 'That guy's huge'." Another former user said, "Steroids were the easy way to get big and to be respected, especially at the gym and they were very easy to obtain.... I see all these people who are built up on TV and stuff and I'm always comparing myself to them all the time, like, 'I'm bigger than him,' or 'I'm more cut up than he is.'" The symptoms of steroid use are different for males and females. In males, steroid use can lead to:

- · Loss of head hair
- · Shrinking of the testicles and lowered sperm count
- Enlargement of the left ventricle of the heart, which can lead to heart problems in later life
- Increased risk of heart attack and stroke
- High blood pressure
- High blood cholesterol levels
- · Weakening of joints and increased risk of rupturing tendons
- Liver damage
- Stunted growth
- Depression, increased aggressiveness, and rapid mood changes

Females who use anabolic steroids may undergo virilization, a process in which they develop certain physical characteristics associated with males:

- Growth of facial hair
- Deepening of the voice
- Irregular menstrual cycles and eventual loss of fertility
- Loss or thinning of scalp hair
- Shrinking of the breasts

Other risks related to steroid use include hepatitis or HIV infection from needle sharing; dangerous side effects from contaminated steroids or fakes (other substances sold as steroids) purchased over the Internet; and legal penalties for purchasing or possessing steroids without a prescription. Anabolic steroids are classified as Schedule III drugs, in the same group as narcotic painkillers and barbiturates. Simple possession of any Schedule III substance without a valid prescription is a federal offense punishable by up to one year in prison and/or a minimum fine of \$1,000.

#### Diagnosis

The diagnosis of steroid use may be based on changes in the person's physical characteristics or behavior. A routine medical checkup may indicate liver damage or high blood pressure. Family members or others may notice oily skin or acne on the user's upper back, needle marks, hair

falling out, jaundice or yellowing of the skin (a sign of liver damage), changes in the person's figure, extreme mood swings, or angry outbursts.

The most common medical test for steroid abuse is a urine sample. Other laboratory tests that the doctor may order are blood tests for blood sugar levels (higher than normal may indicate steroid abuse); cholesterol levels; and liver function.

## Treatment

It is important for someone who has been using steroids to talk to a doctor about quitting rather than simply stopping the drugs. Steroids are powerful hormones and can produce such side effects as nausea, vomiting, joint pain, abdominal cramps, dizziness, and low blood pressure when they are stopped abruptly. In many cases the doctor will work out a schedule to taper the patient's use of steroids gradually. The user should be completely honest with the doctor about the following details:

- All the drugs that were taken and where and how they were purchased. This information is important to help the doctor find out whether the drugs might have been contaminated or counterfeit.
- The user's dosing schedule.
- Length of usage.
- A description of any physical and mental side effects that were experienced.

A person who is coming off steroids should not stop exercising or working out. Physical exercise helps in avoiding depression as well as maintaining good general health and fitness. A balanced diet is also important, as is drinking plenty of water.

In some cases the doctor may recommend psychotherapy to treat the emotional dimension of steroid addiction.

## Prognosis

The prognosis of steroid use depends on the user's gender, length of use, types and amounts of steroids used, and the age at which use began.

## Prevention

The most important preventive measure is good communication between parents and children, particularly during adolescence, when concerns about appearance and achievement become critical to many teens. In many cases athletic coaches or physical education teachers can help by

#### Steroid Use

#### WORDS TO KNOW

**Anabolic:** Referring to tissue building. Anabolic steroids build up muscle and bone tissue.

Androgens: Male sex hormones.

- **Cycling:** Using steroids in periods of several weeks or months (a time cycle) separated by short rest phases of not using the drugs.
- **Hormone:** Any chemical produced by living cells that stimulates organs or tissues in parts of the body at some distance from where it is produced.
- **Stacking:** Using several different types of steroids at the same time.
- **Virilization:** The development of male sexual characteristics in females.

talking frankly about the dangers of steroids to young people who may be experimenting with them.

#### The Future

Further research needs to be done about the long-term effects of anabolic steroids—including their psychological effects—at all stages of the life cycle. Beginning in 2005, a number of steroid activists began to push for decriminalization of anabolic steroids, claiming that they are not as harmful to mature adults as previous studies have indicated. It is difficult to see, however, how adults could be permitted to use steroids freely without the drugs becoming available to teenagers whose bodies are still developing and could be damaged by long-term steroid use.

**SEE ALSO** AIDS; Tendinitis

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# u

## Strabismus

## Definition

Strabismus is a condition in which a person's eyes are not properly aligned with each other. One eye is either constantly or occasionally turned inward, outward, upward, or downward. Someone looking at a person with strabismus may notice that their eyes are pointed in different directions. If one or both eyes are turned inward, the person is said to have crossed eyes; if either eye is turned outward, they are said to be wall-eyed.

Ophthalmologists (doctors who specialize in treating eye disorders) classify strabismus as congenital, acquired, or secondary. Congenital strabismus is present at birth and affects about 1 percent of infants. Acquired strabismus develops in later life from injury to the eye, the brain, or such diseases as diabetes, while secondary strabismus results from another eye disorder, usually cataracts.

#### Description

Strabismus is an eye disorder in which the person's eyes do not focus on a single point at the same time. The lack of coordination of the eyes

## Also Known As

Crossed eyes, squint eye, walleye

#### Cause

Lack or loss of coordination between the two eyes

#### Symptoms

Eye fatigue; seeing double; pulling sensation around the eye; loss of vision in one eye

#### Duration

Lifelong if not corrected

A young boy with strabismus. © IMAGEBROKER / ALAMY.



results from eye muscles that do not work together properly. One or both eyes may turn outward, inward, upward, or downward. Some people may have two different subtypes of strabismus, one in each eye. Strabismus may be constant or it may come and go. In children, strabismus may vary over the course of a few hours as well as from day to day.

Although strabismus is not a life-threatening condition, it can have serious consequences for a child's vision if it is not corrected. The reason is that unlike an adult's fully developed brain, a child's brain is still learning to coordinate the visual perceptions from both eyes. If the two eyes are focusing in different directions, the brain may start to ignore the input from one eye. This condition is called amblyopia, or lazy eye. Although the affected eye is completely normal in terms of its basic structures, it can lose its ability to see over time. Uncorrected amblyopia resulting from strabismus is the leading cause of blindness in one eye in American adults over the age of twenty.

Some children have a condition called pseudostrabismus or false strabismus because their facial features make them look cross-eyed or walleyed. Children with eyes set unusually close together, a wide flat nose, or an extra fold of skin near the inner eye may look as if they have strabismus. False strabismus does not affect vision and usually goes away as the child grows older and his or her face lengthens.

## Demographics

According to the American Academy of Ophthalmology, about four in every one hundred adults in the United States have strabismus. About 2 percent of children have strabismus, half of them being born with the condition. Although strabismus can develop at any age, the age group most commonly affected is children between three and six years of age.

Strabismus appears to run in some families, although no specific genes have yet been identified that are linked to strabismus. It is also not known whether strabismus itself is genetic or whether it is the underlying causes of the disorder (weakness of the eye muscles or cranial nerves controlling the eye muscles) that are inherited. Asian Americans appear to be at a slightly higher risk of strabismus than people of other races or ethnic backgrounds. As far as is known, males and females are equally affected.

Other risk factors for strabismus include: farsightedness or hyperopia; premature birth; and developmental delays.

#### **Causes and Symptoms**

Strabismus in children can be caused by weakness of the eye muscles or defects in the baby's developing nervous system. Children with cere-

## Abraham Lincoln's Strabismus

Abraham Lincoln (1809–1865) was ridiculed by many of his contemporaries for his "homely" and "misshapen" face. The left side of Lincoln's face was noticeably smaller than the right side, a difference that can be clearly seen in photographs taken of him during his presidency. Some medical historians think that this difference was caused by a hereditary developmental defect—Lincoln had a first cousin with strabismus—while others think that the facial problem was caused by trauma. Lincoln had been kicked in the face by a horse when he was nine years old.

Several people who knew Lincoln reported that his left eye sometimes moved upward independently of his right eye—a clear indication of strabismus. Lincoln himself noted that he saw double on occasion; however, he was not particularly troubled by this symptom. In 2007 a retired ophthalmologist and history buff made a laser scan of two facial masks that had been made of Lincoln during his lifetime. The threedimensional scan allowed the doctor to analyze the differences between the muscles in Lincoln's left and right eyes, and to confirm the diagnosis of adult strabismus.

bral palsy, Down syndrome, Edwards syndrome, or whose mother had rubella during pregnancy have a significant risk of congenital strabismus. In adults, acquired strabismus may result from a concussion, brain tumor, stroke, traumatic brain injury that affects the cranial nerve controlling the eye muscles, loss of vision in one eye, or diabetes.

Symptoms of strabismus in a child may include:

- Eyes focusing in different directions when the child becomes interested in playing with a nearby object.
- The child's eyes roll or wander for no apparent reason.
- The child does not seem to have normal depth perception.

• The child covers one eye with the hand in order to focus the other eye.

The symptoms of strabismus in adults are slightly different from those in children because the visual system in the adult's brain is completely developed. Amblyopia is therefore unusual in adults with acquired strabismus. Common symptoms in adults include:

- Seeing double. This is the most annoying symptom for most adults with acquired strabismus because it interferes significantly with reading, driving, and many other everyday tasks.
- Having problems with depth perception.
- Recurrent headaches or feelings of eyestrain.
- Squinting or tilting the head to one side in order to focus the affected eye.
- Eyes that appear crossed or walleyed to other people. This can be a cause of considerable social embarrassment; one study found that 70 percent of adults with strabismus had difficulties in school, work, or family life because of the impact of strabismus on their appearance.

#### Diagnosis

Early diagnosis of strabismus in children is critical in order to prevent amblyopia and later loss of vision. The eyes of newborns cannot always focus directly on an object because the muscles of the eye are still developing. A baby should, however, be able to focus both eyes in parallel by the age of three to four months. If parents notice that the infant still looks cross-eyed or walleyed at four months, they should have the child examined by the child's primary care doctor or by an ophthalmologist.

The most common tests used to diagnose strabismus are the cover test and the Hirschsprung test. In the cover test, the examiner covers each of the patient's eyes in turn with a handheld shield while asking the patient to focus on a small object at a distance. If a person has strabismus, the unaffected eye will not change focus when uncovered, while the strabismic eye will move or change its focus. In the Hirschsprung test, the examiner shines a small flashlight or penlight on the patient's eyes. The examiner can see the light reflected on the front surface of the pupil. If the eyes are properly aligned, the reflection will be in the same spot on each eye. If the person has strabismus, the reflection from the light will appear on different areas of each eye. The ophthalmologist may also perform other examinations to rule out the possibility that a brain tumor is causing the strabismus.

#### Treatment

Treatment of strabismus in children may consist of one or more of the following measures:

- Patch therapy and vision therapy. In patch therapy, the child's stronger eye is covered with a patch, forcing the weaker eye to function at a higher level. Vision therapy consists of exercises performed in the eye doctor's office under supervision to change the way the eyes process visual information. It is not concerned with strengthening the eye muscles.
- Medications injected into the eye muscles to weaken overactive muscles. Interestingly, the drug most often used for this purpose—botox—is derived from the same organism that produces the deadly toxin of botulism food poisoning.
- Corrective lenses. These are intended to improve the focusing of the patient's eyes and redirect the line of sight.
- Surgery. Surgery to realign the eye muscles may be done if corrective lenses and patch therapy are not successful. It is not usually performed on children younger than four years of age.

Adults with strabismus may be treated with corrective lenses, botox injections in the eye muscles, or surgery on the eye muscles. Surgery is usually done to correct double vision, the most troublesome symptom of strabismus in adults.

## Prognosis

When strabismus in children is detected and treated early, the prognosis is good. The prognosis for strabismus in adults depends on the cause (trauma, brain tumor, other diseases) and its severity.

## Prevention

Congenital strabismus can be prevented in some cases by immunization against rubella in women planning a pregnancy. Acquired strabismus can be prevented in some cases by observing safety measures to lower the risk of head injuries in sports or the workplace and by avoiding

#### Strabismus

#### WORDS TO KNOW

**Amblyopia:** Dimness of sight in one eye without any change in the structure of the eye. It is also known as lazy eye.

**Botulism:** A rare but potentially fatal paralytic illness caused by a bacterial toxin in contaminated food. Congenital: Present at birth.

**Pseudostrabismus:** A condition in which a child may seem to have strabismus because of certain facial features that change as the child's face matures.

improperly canned foods (a common source of botulism) or poisonous shellfish.

#### The Future

As of 2008, the National Institutes of Health (NIH) was conducting about 29 different clinical trials related to strabismus. Most are studies comparing the effectiveness of different types of treatment for the condition, but three of the trials are exploring genetic factors that may be linked to strabismus.

**SEE ALSO** Cataracts; Cerebral palsy; Concussion; Diabetes; Edwards syndrome; Hyperopia; Rubella; Stroke

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# Strep Throat

## Definition

Strep throat is an acute infection of the throat caused by the same type of bacterium (group A streptococcus) that causes scarlet fever.

## Description

Strep throat is an infectious disease that may affect adults as well as children. It is spread by fluids from the throat or nose of an infected person or by contact with a drinking glass, soiled tissue, or other object touched by an infected person. In a very few cases, the bacteria that cause strep throat have been spread by contaminated food.

The incubation period of strep throat is short, usually between two and five days. The symptoms often come on rapidly, with the patient developing a sore throat that may make swallowing or speaking difficult. The fever is usually highest on the second day of illness.

## **Demographics**

The bacterium that causes strep throat is common around the world. It is responsible for 25–30 percent of the ten million cases of severe sore throat that doctors treat in the United States each year.

Strep throat can occur at any time of the year but is most common in North America in the late winter and early spring. It is more likely to occur in children between the ages of five and fifteen than in adults; it is less common in children younger than two years of age. Strep throat affects males and females equally, and appears to be equally common in all races and ethnic groups.

## **Causes and Symptoms**

Strep throat is caused by a bacterium—a group A streptococcus that normally lives in the upper respiratory tract. As the bacteria grow in the tissues of the nose and throat, they may secrete toxins that may cause skin rashes in some patients with strep throat in addition to the fever and other symptoms.

#### Also Known As

Streptococcal pharyngitis, streptococcal sore throat

#### Cause

Group A streptococcus; also called *Streptococcus pyogenes* 

#### Symptoms

Sudden and severe sore throat, inflamed tonsils, fever of 101°F (38.3°C), difficulty swallowing

#### Duration

Symptom improvement in two to five days



White areas in the back of this patient's throat signify a strep throat infection. © PHOTOTAKE INC. / ALAMY.

- Abdominal cramps
- Enlarged glands in the neck that are sore to the touch
- Aching muscles or sore joints

#### Diagnosis

Correct diagnosis of strep throat is important because most sore throats are caused by viruses rather than the bacteria that cause strep throat. Sore throat caused by viruses should not be treated with the antibiotics used for strep throat.

The diagnosis of strep throat is usually based on a combination of the patient's history—particularly recent exposure to other children with a streptococcal infection—and the doctor's examination of the patient's throat, neck glands, and skin. The doctor may also take a sample of fluid from the child's throat on a cotton swab. The fluid can be sent to a laboratory for a throat culture, which is an accurate test that takes two days. The doctor may also perform what is called a rapid strep test. It can detect proteins produced by the streptococci in a few minutes in the doctor's office. The rapid strep test, however, is not as accurate as a throat culture.

## Treatment

Patients with a known case of strep throat are treated with a course of antibiotics, most commonly penicillin, azithromycin, or a similar drug known to be effective in treating streptococcal infections. A child whose

- Fever of 101°F (38.3°C) or higher
- Headache
- Chills and cold sweats
- Nausea and vomiting (more common in children)
- Difficulty swallowing
- Red and swollen tonsils, sometimes with yellow or whitish-grey streaks of pus
- Bad breath
- Rash or hives
- Loss of appetite

throat is too sore to allow comfortable swallowing can be given penicillin by injection.

A child with strep throat will usually not be contagious within a day or two of starting treatment and can return to school once the fever goes down. It is important, however, for anyone, child or adult, with strep throat to take the full course of antibiotics (usually ten days), even though he or she may start to feel better in a few days. Not taking the full course of antibiotics may lead to such later complications of strep throat as rheumatic fever or inflammation of the kidneys. The doctor should be consulted if the patient does not begin to feel better after a day or two of antibiotic treatment.

Adults or children with strep throat can take ibuprofen, acetaminophen, or another nonaspirin pain reliever to bring down fever and relieve muscle cramps or headache. Gargling with salt water—one-half teaspoon of salt in a glass of warm water—is recommended for easing the throat discomfort.

In some cases, the doctor may recommend a tonsillectomy to prevent recurrence of streptococcal infections. A study done in 2006 found that children with repeated episodes of strep throat whose tonsils are intact are more than three times as likely to develop additional episodes of strep throat than children whose tonsils were removed.

#### Prognosis

The great majority of patients with strep throat recover completely with no complications even without antibiotic treatment. However, about two patients per 1,000 who are not treated will develop rheumatic fever and another two per 1,000 will develop a severe infection of the tonsils. Other possible but rare complications of strep throat include ear infections, kidney or liver damage, pneumonia, inflammation of the bones or joints, or sinusitis.

Another possible complication of strep throat is the PANDAS syndrome, a term that stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections. Some children develop symptoms of obsessive-compulsive disorder or Tourette syndrome after a group A streptococcal infection; or they become moody, irritable, and anxious when separated from their parents. The connection between these symptoms and the infections is not clear, however, and many doctors think that further research is needed.

#### Prevention

Strep throat is difficult to prevent completely because of the close contact among children in schools and day care centers, particularly in the winter. In addition, some people can carry Group A streptococci in their throats without getting sick themselves. When anyone in a family is diagnosed with strep throat, doctors recommend protecting other members by washing the sick person's eating utensils, drinking glasses, and bedding in hot soapy water separately from the rest of the family's dishes and laundry. Children with strep throat should also be reminded to cover the nose and mouth when sneezing or coughing and to wash the hands thoroughly in warm soapy water afterward.

#### The Future

As of 2008 the chief concern among doctors was the overuse of antibiotics in treating strep throat. Across the United States, 70 percent of children with sore throats who are seen by a doctor are treated with antibiotics even though 30 percent at most have strep infections. It can be difficult to distinguish between bacterial and viral infections of the throat even when a throat culture is performed because some people may be carriers of group A streptococci and have the bacteria in their throat at the same time that they have a sore throat caused by a virus.

The chief danger of overuse of antibiotics is the creation and spread of drug-resistant disease organisms. In addition, some people can have severe allergic reactions to antibiotics. To guard against overprescribing antibiotics for sore throats that may not be caused by streptococci, many doctors now follow a two-step procedure for diagnosing strep throat before giving the patient a prescription for antibiotics. They give the patient a rapid strep test in the office; if the result is positive, the doctor then takes a throat swab and sends it for a culture. If that test too comes back positive, the patient is then given antibiotics. Waiting two days for antibiotic treatment will not increase the patient's risk of developing complications from strep throat.

SEE ALSO Rheumatic fever; Scarlet fever

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## WORDS TO KNOW

**Group A streptococcus:** A sphere-shaped bacterium that grows in long chains and causes strep throat as well as scarlet fever.

**PANDAS disorders:** A group of disorders with psychiatric symptoms that develop in some

children after strep throat or scarlet fever. The acronym stands for Pediatric Autoimmune Neuropsychiatric Disorders Associated with Streptococcal infections.

Pharyngitis: The medical term for sore throat.

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# m

# Stress

## Definition

Stress was originally defined by a Canadian biologist named Hans Selye (1907–1982) in the 1930s as a series of biochemical, nervous, digestive, and muscular responses in laboratory animals to a perceived threat, which he called a stressor. Selye later expanded the notion of stress to include humans trying to evaluate and respond to various stressors in their daily lives. Many people, however, use the word "stress" in casual

conversation to refer to almost any event or situation that worries or upsets them.

Some doctors distinguish several different types of stress according to whether the stress is positive or negative, or according to the length of time that the person experiences the stressor. In the 1970s, a psychologist named Richard Lazarus coined the term *eustress* to refer to stress that is good for health or leads to a sense of fulfillment. Increasing one's strength or endurance through active exercise would be an example of eustress, as would completing a course of study or finishing a job assignment.

Stress can also be categorized as acute, episodic acute, or chronic, according to the length of time the person feels stressed and whether the stress is a one-time event, repeated periodically, or long-term and unrelenting. An example of acute stress is facing a growling and aggressivelooking dog. Episodic acute stress includes the pressures that people place on themselves by taking on too many obligations or by not allowing enough time to complete tasks. Some jobs, such as police work or firefighting, involve episodic acute stress. Chronic stress is usually situational—the person is trapped in a bad marriage, a dead-end job, or living in a crime-ridden neighborhood.

#### Description

In general, stress is a complicated set of physical and emotional responses to changes that occur in everyone's life. On the biological level, stress begins with the "fight-or-flight" reaction—the activation of a section of the brain called the hypothalamic-pituitary-adrenal system, or HPA. When a human perceives a situation as stressful, the HPA system releases cortisol, a steroid hormone. The next stage in the stress reaction is the release of neurotransmitters, or brain chemicals, that activate parts of the brain that register the emotion of fear. The neurotransmitters also suppress activity in parts of the brain associated with short-term memory, concentration, and rational thinking. This limitation allows a human to react quickly to a stressful situation but it also lowers his or her ability to deal with intellectual or social factors that may be part of the situation.

On the physical level, the person's heart rate and blood pressure rise; he or she breathes more rapidly, which allows the lungs to take in more oxygen. Blood flow to the muscles, lungs, and brain may increase by 300–400 percent. The spleen releases more blood cells into the

#### Also Known As

Psychological stress, anxiety, tension

#### Cause

The body's responses to any event or situation perceived as a stressor

#### Symptoms

Anxiety, trouble sleeping, moodiness, nausea, depression, overeating, drinking or doing drugs

#### Duration

Minutes to years

**Stress** 

circulation, which increases the blood's ability to transport oxygen. The immune system redirects white blood cells to the skin, bone marrow, and lymph nodes. At the same time, nonessential body systems shut down. The skin becomes cool and sweaty as blood is drawn away from it toward the heart and muscles. The mouth becomes dry, and the digestive system slows down.

After the crisis passes, the levels of stress hormones drop and the body's various organ systems return to normal. This return is called the relaxation response. Some people are more vulnerable to stress than others because their hormone levels do not return to normal after a stressful event. In chronic stress, the organ systems of the body do not have the opportunity to return fully to normal levels. Different organs become under- or overactivated on a long-term basis. In time, these abnormal levels of activity can damage an organ or organ system.

What complicates the experience of stress is that different people respond differently to stressors. Some people find driving a highly stressful experience, for example, while others enjoy it. Similarly, some people enjoy the intellectual challenges of certain fields of study, while others are bored by them. Personality differences are another factor that influences people's response to stress. Some people are highly aggressive, inclined to worry, or easily irritated, while others are less competitive or more optimistic about life.

#### Demographics

Almost everyone has experienced acute or episodic acute stressors. Some people, however, are more vulnerable than others to chronic stressrelated illnesses:

- Children. Children have very little control over their environments. In addition, they are often unable to communicate their feelings accurately.
- Elderly adults. Aging appears to affect the body's response to stress, so that the relaxation response following a stressful event is slower and less complete. In addition, the elderly are often affected by such major stressors as health problems, the death of a spouse or close friends, and financial worries.
- Caregivers of mentally or physically disabled family members.
- Women in general.
- People with less education.

- People who belong to racial or ethnic groups that suffer discrimination.
- People who live in cities.
- People who are anger-prone. Chronic anger is associated with narrowing of the arteries, a factor in heart disease.
- · People who lack supportive relatives or friends.

#### **Causes and Symptoms**

Stress is caused by the human body's response to any event or situation perceived as a stressor. Perception is an important factor in a person's reaction to stress because it can be modified or changed in some situations.

The specific symptoms of stress-related illness vary from person to person depending on which organs or body systems are most vulnerable. Common symptoms of stress include:

- Heart. Chronic stress raises blood pressure, triggers the release of cholesterol into the bloodstream, and causes the arteries to narrow. It also increases the possibility that a clot will form in the coronary arteries, thus increasing the person's risk of heart attack or stroke.
- Skin. Eczema and other allergic skin rashes can be triggered or made worse by stress.
- Digestive tract. Stress leads to nausea, diarrhea, constipation, bloating, and irritable bowel syndrome in many people. It may also play a role in the onset of eating disorders.
- Reproductive system. Stress can lead to loss of sexual desire in both men and women. In addition, stress during pregnancy is associated with a 50 percent higher risk of miscarriage. High stress levels on the mother during pregnancy are also related to higher rates of premature births and babies of lower than average birth weight; both are risk factors for infant mortality.
- Bones, joints, and muscles. Stress intensifies the chronic pain of arthritis and other joint disorders. It also produces tension-type headaches, which are headaches caused by the tightening of the muscles in the neck and scalp.
- Brain and central nervous system. Stress hormones released during acute stress interfere with memory and learning. People who are under severe stress become unable to concentrate; they may

become clumsy and accident-prone. Acute stress interferes with short-term memory, although this effect goes away after the stress is resolved. In children, however, the brain's biochemical responses to stress clearly limit the ability to learn.

• Immune system. Chronic stress increases a person's risk of getting an infectious illness. Several research studies have shown that people under chronic stress have lower than normal white blood cell counts and are more vulnerable to colds and influenza. Men with HIV infection and high stress levels progress more rapidly to AIDS than infected men with lower stress levels.

#### Diagnosis

There is no specific test or imaging study for diagnosing stress. People with specific mental disorders like posttraumatic stress disorder (PTSD) or panic disorder can be diagnosed by a psychiatrist qualified to evaluate these conditions. In most other cases, however, a person is diagnosed in the course of a checkup for a stress-related physical condition when the doctor asks about the stress level in their living situation, school, or job.

## Treatment

Treatment for stress depends on the parts of the person's body that are affected and the sources of stress and types of stress in his or her life. Most people benefit from a combination of treatment approaches:

- Medications. These can be prescribed to treat physical conditions related to stress like high blood pressure or high cholesterol levels, or to help relieve emotional anxiety.
- Psychotherapy. The two approaches most often used in treating stress are interpersonal therapy and cognitive therapy. In interpersonal therapy, people learn about the events in their past and the triggers in their lives that set off the stress response, together with strategies for coping with stressors. Cognitive therapy works by teaching patients to change their ways of thinking about stressful situations or events. Many people have underlying negative assumptions about life that make them more vulnerable to stress.
- Lifestyle changes. People who have used alcohol, drugs, or smoking are usually advised to quit. In some cases, people may have to change jobs or leave bad relationships in order to relieve chronic stress.

- Physical exercise. Physical activity is a good way to work off tension in the muscles and joints, and to improve strength and endurance.
- Stress management. Stress management refers to programs or techniques intended to help people deal more effectively with stress. Many of these are intended to help people handle job- or workplace-related stress. Stress management programs ask participants to identify the specific aspects of their jobs that they find stressful and then plan a course of positive action to lower their stress levels.
- Complementary and alternative approaches. Acupuncture, yoga, relaxation training, meditation, prayer and religious practice, guided imagery, hypnosis, massage therapy, music therapy, humor, and pet therapy are alternative approaches that help many people cope with stress.

## Prognosis

The prognosis for stress depends on the patients' overall health, their age, the specific stressors they confront, and the ways of coping that they have developed over time. Some people tend to focus on their feelings about the stressful situation while others focus on solving the problem. Still others react to stress by trying to escape from it through drugs or alcohol.

## Prevention

The National Institutes of Health (NIH) has compiled a list of ten things people can do to prevent stress:

- Stay away from stressors that can be avoided.
- Avoid making too many lifestyle changes too close together—such as trying to quit smoking while planning to move to a new city and take a new job.
- Recognize limitations and not take on too many responsibilities at the same time.
- Organize tasks according to priority and allow enough time to complete each one.
- Learn to communicate effectively and politely with others.
- Don't isolate; share thoughts or feelings with friends, family, or appropriate others, and take their advice if it seems reasonable and helpful.

## WORDS TO KNOW

**Eustress:** A term that is sometimes used to refer to positive stress.

**Stress management:** Any set of techniques intended to help people deal more effectively with stress in their lives by analyzing specific stressors and taking positive actions to minimize their effects.

**Stressor:** Any event or stimulus that provokes a stress response in a human or animal.

- Create a positive attitude toward life.
- Set aside time for a break or a treat as a reward for overcoming a stressful situation.
- Get regular physical exercise, at least thirty minutes each day.
- Eat a healthy diet and get enough sleep.

## The Future

The stress level in people's lives is likely to increase in the future rather than improve. One reason is the growing complexity and interconnectedness of the world; political, economic, and public health problems in almost any country can have an impact on others thousands of miles away. Another factor is the effect of the mass media and rapid communications. People are constantly bombarded with news about natural disasters and other frightening events.

**SEE ALSO** Depression; Eczema; Heart attack; Irritable bowel syndrome; Panic disorder; Posttraumatic stress disorder; Prematurity; Stroke

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# Stroke

## Definition

Stroke is a life-threatening condition that occurs when the blood supply to a part of the brain is suddenly cut off or when brain tissue is damaged by bleeding into the brain. There are two main types of stroke. Ischemic stroke occurs when a clot formed in the artery or coming from elsewhere in the artery system blocks an artery to the brain; this type accounts for about 80 percent of strokes. The other type, hemorrhagic stroke, occurs when a blood vessel in the brain bursts, allowing blood to spill out into brain tissue. The blood upsets the chemical balance that the nerve cells in the brain need to function.

## Description

Stroke is usually a sudden occurrence. Some people have a warning event called a transient ischemic attack (TIA) or mini-stroke. A TIA has the

Also Known As Apoplexy, brain attack, cerebrovascular accident

#### Cause

Shortage of blood supply to the brain

#### Symptoms

Weakness or paralysis on one side, headache, vision problems, confusion, difficulty speaking

#### Duration

Brain damage begins within minutes; may continue for days

#### Stroke



*Elderly woman undergoing rebabilitation after a stroke.* © DENNIS MACDONALD / ALAMY.

same symptoms as a full-blown stroke but goes away in a few minutes or hours, leaving no permanent effects. It is, however, an indication that the person is at risk of a major stroke and should see their doctor right away. A TIA offers the person an opportunity to take preventive action.

Stroke has five major signs or symptoms. The American Stroke Association has a quick symptom checklist called "Give Me 5"

- Walk: Is the person having trouble with balance or coordination?
- Talk: Is speech difficult or slurred? Is the person's face drooping?
- Reach: Is one side of the body weak or numb?
- See: Is vision partly or entirely lost?
- Feel: Does the person have a sudden severe headache with no obvious cause?

A person with stroke can have more than one of these symptoms at the same time. The important feature to keep in mind is that the symptoms come on suddenly, which helps in distinguishing stroke from other causes of dizziness, vision problems, or headache.

## **Demographics**

According to the Centers for Disease Control and Prevention (CDC), stroke is the third leading cause of death in the United States as of 2008, being responsible for about 160,000 deaths each year. About 700,000

## **First Aid for Stroke**

The most important first step is to call for emergency help *at once*. Stroke is a medical emergency; the sooner the person is evaluated and treated, the better their chances of recovery. The drug presently considered most useful in treating stroke must be given within three hours of the attack to be effective.

Additional measures that can be taken while waiting for the emergency team:

- If the person stops breathing, give them mouth-to-mouth resuscitation.
- If they are vomiting, tilt their head to one side to prevent them from swallowing the material.
- Do *not* give them anything to eat or drink.

Americans have strokes each year, 500,000 for the first time and 200,000 having a second or third stroke. The total cost of stroke to the American economy each year is about \$43 billion.

About 50,000 Americans have a TIA in an average year; of this group, 35 percent will have a severe stroke at some point in the future.

Strokes can affect people in any age group; however, the risk increases sharply in people over fiftyfive years of age. Seventy-five percent of all strokes occur in people over sixty-four. Men are 1.25 times more likely to have strokes than women; however, women are more likely to die of stroke because they are usually older when they have their first stroke.

African Americans have an increased risk of stroke compared to other racial and ethnic groups in the United States, and they are also more likely to suffer a stroke at younger ages. African Americans between the ages of forty-five and fifty-five die from stroke 4–5 times more often that Caucasians in the same age group.

Risk factors for stroke include:

- Hypertension (high blood pressure). This is the most important single risk factor for stroke.
- High blood cholesterol levels.
- Being over age fifty-five.
- A family history of stroke, TIA, or heart attack.
- Diabetes.
- Smoking. Smoking doubles a person's risk of ischemic stroke.
- Personal history of previous stroke or TIA.
- Obesity.
- Heavy use of cocaine.
- Irregular heart rhythm.
- Heavy drinking. Alcohol consumption raises a person's blood pressure.
- Use of birth control pills or hormone replacement therapy.

#### **Causes and Symptoms**

Stroke is caused by a loss of blood supply to the brain resulting either from a clot blocking an artery or from bleeding into or around the brain. Ischemic stroke can result from two types of clots. The first is an embolus, which is a free-floating clot produced in the heart or somewhere else in the body that travels to a blood vessel in the brain. The second type of clot is formed within an artery in the head or neck and grows there until it is large enough to block the artery. Atherosclerosis, a disease of the blood vessels in which fatty deposits build up along the walls of the vessels, is a common cause of this type of clot.

Hemorrhagic stroke can occur when an aneurysm—a weak spot in the wall of an artery—suddenly bursts. High blood pressure is the most common cause of this type of hemorrhagic stroke. Hemorrhagic stroke can also occur when the walls of an artery become thin and brittle; they can then break and leak blood into the brain. Hemorrhagic stroke can take one of two forms: the blood can leak directly into brain tissue from an artery in the brain, or it can leak from an artery near the surface of the brain into the space between the skull and the membranes covering the brain.

The major symptoms of stroke have already been described. Other symptoms that some patients experience include drooling, uncontrollable eye movements, personality or mood changes, drowsiness, loss of memory, or loss of consciousness.

#### Diagnosis

The diagnosis of stroke includes taking the patient's history and obtaining an account of the patient's symptoms, followed by a complete physical and a neurological examination to rule out the possibility that the patient's symptoms are being caused by a brain tumor. The neurologist may use the National Institutes of Health Stroke Scale (NIHSS), which is a checklist that allows the doctor to record the patient's level of consciousness; visual function; ability to move; ability to feel sensations; ability to move the facial muscles; and ability to talk. Other tests include:

- Blood tests. These can reveal the existence of blood disorders that increase a person's risk of stroke.
- Computed tomography (CT) scan. This type of imaging test is one of the first tests given to a patient suspected of having a stroke. It

helps the doctor determine the cause of the stroke and the extent of brain injury.

- Magnetic resonance imaging (MRI). This imaging test is useful in pinpointing the location of small or deep brain injuries.
- Electroencephalogram (EEG). This test measures the brain's electrical activity.
- Blood flow tests. These are done to detect the location and size of any blockages in the blood vessels. One type of blood flow test uses ultrasound to produce an image of the arteries in the neck leading into the brain. Another type of blood flow test, called angiography, uses a special dye injected into blood vessels that will show up on an x ray.
- Echocardiography. This type of test uses ultrasound to produce an image of the heart. It can be useful in determining whether an embolus from the heart caused the patient's stroke.

## Treatment

Treatment of stroke depends on whether it is ischemic or hemorrhagic. Ischemic stroke is treated first with blood thinners, often aspirin or another drug known as warfarin. If the patient is seen by a specialized stroke team within three hours of the attack, he or she may be treated with a drug called tissue plasminogen activator or tPA. It is critical, however, to be sure that the patient has an ischemic rather than a hemorrhagic stroke, as blood-thinning drugs can make a hemorrhagic stroke worse.

Hemorrhagic stroke is treated by removing pooled blood from the brain and repairing damaged blood vessels. To prevent another hemorrhagic stroke, the surgeon may use a procedure called aneurysm clipping. In this procedure, the surgeon clamps the weak spot in the artery away from the rest of the blood vessel, which reduces the chances that it will burst and bleed.

After emergency treatment in the hospital, most stroke patients need long-term recovery and rehabilitation. The type and length of therapy depend on the amount of function the patient has lost; some need to relearn language skills, while others may need to relearn bowel and bladder control, swallowing, and movement or balance. Most patients need a combination of physical therapy, occupational therapy, speech therapy, and psychotherapy. It is common for stroke patients to feel depressed during rehabilitation or to have trouble controlling their feelings.

## Prognosis

The prognosis of stroke depends on the person's age, the type and location of the stroke, and the amount of time elapsed between diagnosis and treatment. In general, patients with ischemic stroke have a better prognosis than those with hemorrhagic stroke. In one study in the Boston area, 19 percent of patients with ischemic stroke died within the first thirty days of the attack compared to 35 percent with hemorrhagic stroke.

About 10 percent of stroke patients recover enough function to live independently without help; another 50 percent can remain at home with outside assistance. The remaining 40 percent require long-term care in a nursing home.

## Prevention

People cannot change some risk factors for stroke, such as race, age, sex, or family history, but they can control several other risk factors:

- They can quit smoking, drinking heavily, or using cocaine.
- They can keep their weight at a healthy level.
- They can exercise regularly, eat a healthy diet, and take medications for high blood pressure if they are diagnosed with it.
- They can take steps to lower their risk of diabetes or high blood cholesterol levels.
- They can lower the level of emotional stress in their life or learn to manage stress more effectively.
- They can get regular checkups for abnormal heart rhythms if they have been diagnosed with such problems.
- They can see their doctor at once if they have a TIA.

## The Future

Stroke is a disorder that has attracted researchers from a number of different fields because its costs to individuals are still high and doctors are increasingly recognizing that many strokes are preventable. In addition, the aging of the American population means that the number of stroke patients is likely to increase over the next several decades. As of 2008, the National Institutes of Health was sponsoring 1,800 separate studies of stroke prevention and treatment, ranging from new medications to treat
#### Stroke

#### WORDS TO KNOW

- **Aneurysm:** A weak or thin spot on the wall of an artery.
- **Embolus:** The medical term for a clot that forms in the heart and travels through the circulatory system to another part of the body.
- **Ischemia:** Loss of blood supply to a tissue or organ resulting from the blockage of a blood vessel.
- **Thrombus:** A blood clot that forms inside an intact blood vessel and remains there.
- **Transient ischemic attack (TIA):** A brief stroke lasting from a few minutes to twenty-four hours. TIAs are sometimes called mini-strokes.

ischemic stroke to investigations of the genetic factors that increase people's risk of stroke.

A recent innovation is the use of computer technology to allow stroke experts in one hospital to evaluate and diagnose a patient in another hospital that might not have a specialist available. Called Tele-Stroke, the network allows a patient to be evaluated for ischemic stroke within the three-hour time limit for the effective use of tPA.

**SEE ALSO** Coronary artery disease; Hypercholesterolemia; Hypertension; Sickle cell anemia; Smoking

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# U

# Sudden Infant Death Syndrome

# Definition

Sudden infant death syndrome, or SIDS, is the unexpected death of an apparently healthy baby. According to the Centers for Disease Control and Prevention (CDC), such a death "cannot be explained after a thorough investigation is conducted, including a complete autopsy, examination of the death scene, and review of the baby's clinical history."

# Description

In the typical SIDS case, the parents or caregivers put the baby to bed after feeding him or her. A check of the baby shortly after bedtime indicates that everything is normal. However, the baby is later found dead, usually in the position in which he or she had been placed at bedtime or naptime.

In most cases of SIDS, the parents state that the child was apparently healthy. However, some parents of infants who died of SIDS state that their babies "were not themselves" in the hours before death. In a number of cases, the parents report that the baby had diarrhea and vomiting at some point in the two weeks prior to death. Currently, doctors do not know whether these digestive problems are related to SIDS in some way or are only coincidental.

# Demographics

According to the CDC, SIDS is the leading cause of death among American infants between the ages of one and twelve months, and is the third leading cause overall of infant mortality in the United States. SIDS is Also Known As SIDS, crib death, cot death, sudden unexpected death in infancy

**Cause** Unknown

#### Symptoms

Infant found dead after having gone to sleep in apparent normal health

Duration

A few hours

# "Back to Sleep" Campaign

The "Back to Sleep" Campaign is a safety program promoted by the National Institute of Child Health and Human Development (NICHD) that recommends placing a baby on its back for naps and sleeping at night. The campaign was started in 1992 following a recommendation from the American Academy of Pediatrics that babies should not be placed on their stomachs to sleep—as had been common practice for many years.

The Back to Sleep campaign emphasizes three central points:

- Sleeping on the back reduces an infant's risk of sudden infant death syndrome (SIDS).
- The sleeping surface makes a difference. Babies who are put to bed on a soft surface are at increased risk of SIDS.
- Every sleep and nap time matters. Babies should *always* be placed on the back for sleeping. According to the campaign, "Babies who usually sleep on their backs but who are then placed on their stomachs are at very high risk for SIDS."

responsible for about one death per 2,000 live births as of the early 2000s; however, this figure is more than 50 percent lower than the figures for 1990, largely as a result of the "Back to Sleep" campaign.

Most SIDS deaths occur in babies between two and four months of age; only 1 percent occur in newborns. Boys are more likely than girls to die of SIDS; 60–70 percent of SIDS cases involve boys.

According to NICHD, African American babies are twice as likely as Caucasian babies to die from SIDS, and Native American babies are three times as likely. The reason for these differences is not yet known but may be related to other risk factors listed below.

Studies indicate that some mothers are at increased risk of having their child die of SIDS:

- Those who smoke during pregnancy and after childbirth.
- Those who abuse drugs or alcohol.
- Those who are underweight or suffer from malnutrition.
- Those who have children less than one year apart.
- Teenage mothers. The more children the mother has while still in her teens, the greater the risk of SIDS.
- Those who are obese.

Apart from sleeping position, some babies are at increased risk of SIDS:

- Babies who are born prematurely.
- Babies who weigh 4 pounds (1800 grams) or less at birth.
- Babies who are not breastfed.
- Babies who are part of a set of twins, triplets, or quadruplets.
- Babies who are exposed to tobacco smoke.
- Babies put to sleep in an overheated room.

- Babies whose parents practice co-sleeping (the baby shares the parents' bed at night).
- Babies who are overdressed for sleep or covered with too many blankets.

# **Causes and Symptoms**

The cause of SIDS is not known with certainty. It is possible that some cases of SIDS are the result of a combination of factors. Doctors have proposed several different theories for SIDS:

- Bacterial infections. A British study published in May 2008 reported that some cases of SIDS appear to result from previously undetected bacterial infections.
- Abnormalities in the part of the brain stem that controls breathing. A study published in the *Journal of the American Medical Associa-tion* in the fall of 2007 is one of the strongest pieces of evidence so far that innate differences in brain structure may put some babies at increased risk of SIDS.
- Smothering caused by sleeping on the stomach. This theory holds that babies put to sleep lying on the stomach may breathe in their own exhaled carbon dioxide because they do not have the same ability as older children to move their heads during sleep to get more oxygen.
- Episodes of apnea (sudden cessation of breathing). Babies sometimes stop breathing periodically for reasons that are still not understood.
- Abnormalities in heart rhythm. About 10 percent of babies who die of SIDS have been found to have a gene associated with sudden episodes of extremely rapid heartbeat.
- Triple-risk theory. This theory proposes to explain SIDS as the end result of three factors: a biological vulnerability (such as a weakened heart or abnormal brain stem), an environmental problem (such as sleeping on the stomach), and being too young to regulate breathing and other vital functions as effectively as older children.

Theories that are no longer accepted include the notion that SIDS is caused by vaccinations, by dust mites or other insects in the crib mattress, or by toxic gases released by materials used in the manufacture of crib mattresses.

The symptoms of SIDS have already been described.

#### Diagnosis

SIDS is a diagnosis of exclusion, which means that the doctor can list it as the cause of a baby's death only after all other possible causes have been ruled out. It is particularly important to exclude the possibility that the baby had been abused. The American Academy of Pediatrics has drawn up a list of criteria that must be met in order to distinguish SIDS from child abuse:

- There has been a complete autopsy of the baby, and the autopsy findings are consistent with a diagnosis of SIDS.
- There is no evidence of head trauma or significant disease.
- There is no evidence of trauma to the baby's bones.
- Other possible causes of death have been ruled out, including pneumonia, metabolic disorders, dehydration, severe birth defects, dehydration, massive infection, trauma to the abdomen, or carbon monoxide poisoning.
- There is no evidence that the baby was given alcohol, drugs, or other toxic substances.
- There is no evidence of foul play when the death scene is investigated.
- The baby's medical history does not indicate previous health problems.

# Treatment

There is nothing that can be done to treat the infant when SIDS occurs. Treatment of the parents includes support and understanding. However, the doctor and other health professionals must at the same time conduct a thorough investigation into the circumstances surrounding the baby's death. There are some differences among the states as to the way in which the postmortem (after death) investigation is carried out, but all states require an investigation before the death can be defined as SIDS. It is difficult for many parents to accept the need for an autopsy and an evaluation of the bed and room in which the baby died when they are grieving. However, ruling out the possibility of abuse or intentional suffocation of the child is a legal necessity.

Circumstances that concern doctors as well as law enforcement when a baby dies suddenly include:

- The child was seven months of age or older. SIDS is unusual in this age group.
- The pregnancy was unwanted.
- There have been previous unexplained infant deaths in the family.
- Family members have a history of arrests for violent behavior.

# Prevention

The CDC recommends the following precautions to reduce the risk of SIDS:

- Infants should always be placed on their backs to sleep when they are left alone; they should be placed on their stomachs *only* when they are awake and supervised by someone responsible.
- If the baby sleeps in a crib, the crib's mattress should be firm and fit snugly into the crib frame. Such other firm sleeping surfaces as bassinets or cradles are also fine.
- The baby should be dressed in a sleeper or pajama to keep it warm rather than being covered by a blanket.
- Parents who co-sleep with a baby should *never* smoke, drink alcohol, or use drugs when sleeping with the baby. It is better to have the baby sleep in a crib or bassinet next to the parents' bed rather than sharing the bed.
- Parents should *never* put a baby on a couch, waterbed, or pillow for a nap.
- Parents should *never* smoke in the same room as the baby or allow anyone else to do so.
- Caregivers should *never* place the baby to sleep or nap with any pillows, stuffed toys, bumper pads, comforters, quilts, or sheepskins.

# The Future

Present research into SIDS is focused on obtaining a better understanding of the causes of the tragedy, and the factors that may increase

#### WORDS TO KNOW

**Autopsy:** The examination of a body after death to determine the cause of death.

**Brain stem:** The lower part of the brain directly connected to the spinal cord. It controls breathing and other vital functions.

**Co-sleeping:** Allowing a baby to sleep in the same bed as its parents. It is also called bed sharing.

**Postmortem:** Referring to the period following death.

a child's risk of dying of SIDS. Some scientists are looking into possible genetic factors that may affect a child's risk of SIDS.

**SEE ALSO** Child abuse; Obesity; Prematurity; Shaken baby syndrome; Smoking

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# Sunburn

# Definition

Sunburn is an inflammation of the skin caused by overexposure to ultraviolet (UV) radiation, usually from the sun.

# Description

Sunburn is an uncomfortable skin condition marked by reddening and soreness in its milder forms and peeling or blistering with longer periods of exposure to sunlight or tanning lamps. The skin is hot and painful to the touch. Sunburn can also affect the eyes, causing a dry or gritty feeling inside the eyelid.

# Demographics

Sunburn is very common in the general population in North America. According to a survey carried out by the Skin Cancer Foundation, 42 percent of the people who answered the survey reported getting sunburned at least once in the preceding year.

Some people are at greater risk of sunburn than others. Risk factors include:

- Fair skin. Fair-skinned people with red or blond hair and lightcolored eyes are at particularly high risk of sunburn.
- Infants and children of all races.
- People with diabetes or thyroid disease.
- People who live at high altitudes or close to the tropics, and people who are traveling to those parts of the world.
- People who take certain types of antibiotics, tranquilizers, or birth control pills; these drugs make the skin more sensitive to sunlight.
- People who enjoy swimming or boating in the summer or skiing in the winter. Sunlight reflected from water or snow can damage the skin as well as sunlight falling directly on the skin.
- People whose work requires them to spend a lot of time in the sun.

Also Known As Sun overexposure, photodermatitis

#### Cause

Exposure to ultraviolet rays from sun or tanning devices

#### Symptoms

Reddened skin painful to touch, blisters, skin peeling

#### Duration

A week to several weeks following exposure

*Severe blister due to sunburn.* © TONY WATSON / ALAMY



#### **Causes and Symptoms**

Sunburn is caused by ultraviolet (UV) radiation from the sun or a tanning lamp. The UV radiation causes damage to the DNA (genetic material) in the cells of the skin. The body responds to this damage by repairing the DNA and by increasing production of melanin, a brownish-black pigment that protects the skin from further sun damage. Melanin is responsible for the changes in skin color known as a suntan.

Over time, repeated episodes of sunburn can lead to dry skin, premature wrinkling of the skin, patches of scaly skin known as actinic keratosis, and various types of skin cancer. Repeated exposure to sunlight over a period of years can also lead to changes in the lens of the eye known as a cataract.

The symptoms of mild sunburn are:

- Redness and pain in the affected skin. The intensity of the pain is directly proportional to the length of time the person was exposed to UV radiation and the intensity of the radiation. A person can get sunburned in as little as fifteen minutes. The redness usually develops within thiry minutes to six hours after exposure. The pain is most intense from six to forty-eight hours after exposure.
- The skin becomes swollen.

- The skin becomes itchy.
- The skin is hot to the touch. This warmth is caused by the dilation of blood vessels in the injured area.
- The area may exhibit peeling after several days.

More severe sunburn causes the formation of blisters on the affected skin. Skin usually begins to peel about three days after exposure and may continue for another week or ten days.

People who are severely sunburned sometimes develop a condition called sun poisoning. The symptoms of sun poisoning include fever, chills, dizziness, fluid loss, extreme tiredness, and a skin rash as well as the sunburn.

# Diagnosis

Most people do not need to consult a doctor to diagnose sunburn. Mild sunburn can be treated at home (see sidebar). However, people who notice any of the following symptoms should see a dermatologist (a doctor who specializes in skin problems) because they may be early signs of skin cancer:

- A growth on the skin that was not there before the sunburn.
- A sore that bleeds, crusts over, keeps reopening, and does not heal within two weeks.
- A change in the size, color, or texture of a mole.
- A dark flat spot on the skin that is gradually enlarging.

# Treatment

Home treatment of mild sunburn is described in the sidebar. Most mild cases of sunburn

# First Aid for Sunburn

The discomfort of sunburn can be relieved at home by the following treatments:

- Taking a cool (not cold) bath or shower, adding baking soda to the water if desired.
- Soothing the skin with a washcloth soaked in cold skim milk. The skim milk contains protein that helps to ease the pain.
- Applying a non-greasy lotion moisturizing lotion to the burned area. Products containing aloe vera gel are a good choice.
- A non-aspirin pain reliever like Advil or Motrin helps relieve the irritation.

There are also some home remedies and cosmetic cover-ups that should be avoided:

- Do not apply a self-tanning lotion to sunburned or peeling skin; it will stick to the injured skin and make it look worse.
- Do not use petroleum jelly, butter, or products containing local anesthetics to the burned area. They will make the discomfort worse and slow healing.
- Do not use harsh soap to wash sunburned skin.
- If blisters form, do not cover them with ointments; allow them to heal in the open air. If they break open to form sores, cover them lightly with clean gauze and apply an antibacterial lotion to prevent infection.
- Do not give aspirin to children or teenagers to relieve the inflammation of sunburned skin; use a non-aspirin pain reliever instead.

eventually heal without special attention from a doctor. Blisters, however, may require medical treatment if they break open and become infected. A person with sunburn blisters that are oozing, hot, red, swollen, and painful should see their doctor.

A patient with any of the following symptoms may be suffering from sun poisoning, heat exhaustion, or shock as well as sunburn. A doctor should be contacted at once:

- Dizziness or a faint feeling
- A fast pulse or rapid breathing
- Sunken eyes, no urine output, extreme thirst
- Pale, cool, or clammy skin
- Severe or painful blisters
- Eyes hurt or are sensitive to light
- Nausea, fever, chills, or a skin rash

#### Prognosis

Most mild cases of sunburn heal without problems in the short term. Blisters that become infected usually heal completely once the infection is treated. The long-term prognosis is of greater concern, as a history of repeated sunburn increases a person's risk of melanoma (the most serious form of skin cancer) as well as cataracts and other eye disorders.

#### Prevention

Sunburn can be prevented by taking the following steps:

- Avoiding tanning booths and sun lamps.
- Staying out of the sun between 10 a.m. and 4 p.m.
- Using a sunscreen with a sun protection factor (SPF) of 15 or higher every day. People with very fair skin should use a product with an SPF of 30 or higher.
- Applying sunscreen over the entire body thirty minutes before going outside, and reapplying the product every two hours.
- Using a lip balm that contains sunscreen.
- Wearing clothing that covers as much of the body as possible, including a broad-brimmed hat and sunglasses to protect the eyes.
- Keeping infants under six months out of the sun altogether, and using sunscreen on infants older than six months.

# WORDS TO KNOW

Actinic keratosis: A patch of thickened or scaly skin caused by sun exposure. It is not itself a form of skin cancer but may develop progressively into a skin cancer.

**Dermatology:** The branch of medicine that deals with skin problems and disorders.

Melanin: A brownish-black skin pigment.

- **Melanoma:** The most serious form of skin cancer. Sunburn increases the risk of melanoma.
- **Sun poisoning:** A term sometimes used to refer to a severe reaction to sunburn, consisting of fever, chills, fluid loss, dizziness, and nausea.

#### The Future

Sunburn is a common health problem that is not likely to go away any time soon. One reason is that it can easily happen accidentally to someone who may not have planned to be outside in the sun for more than a few minutes. Another reason is that the long-term risks of sunburn, such as skin cancer and cataracts, usually take years to appear. This time lag means that a teenager who wants a glamorous tan right now may not think much about what will happen to his or her skin twenty or thirty years in the future. Prevention of sunburn is an important health measure for people of all ages.

SEE ALSO Burns and scalds; Cataracts; Dermatitis; Heat exhaustion

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# Syphilis

# Definition

Syphilis is a sexually transmitted disease (STD) caused by a spiral-shaped bacterium, *Treponema pallidum*. It can be transmitted by an infected mother to her unborn child as well as by genital, oral, or anal sexual contact. Although the disease has been known for centuries, the organism that causes it was not identified until 1913. The discoverer was Hideyo Noguchi (1878–1928), a Japanese bacteriologist who was working at the Rockefeller Institute in New York as a research assistant.

# Description

Syphilis was for many years one of the deadliest STDs, not only because there was no effective treatment for it before the twentieth century, but also because its symptoms are difficult to distinguish from those of other diseases. Syphilis was nicknamed the "Great Imitator" because it could be easily confused with leprosy, infectious mononucleosis, fungal infections of the skin, meningitis, genital herpes, lymphoma, and many other diseases.

To add to the diagnostic confusion, the symptoms of syphilis have changed somewhat since the first recorded European outbreak in 1494. French troops fighting in Italy were stricken with an STD that caused death within a few months. Medical historians think that this first epidemic, called the great pox to distinguish it from smallpox, was so severe because the disease was either new to Europe or that the spirochete that causes syphilis had mutated over the centuries. Some researchers hold that syphilis was brought to Europe by Columbus's sailors, while others maintain that skeletons from the Middle Ages found in both Italy and England show evidence of late-stage syphilis. In either case, syphilis as it is known in the twenty-first century is rarely fatal unless it is not treated.

#### Also Known As

*Treponema pallidum* infection, lues

#### Cause

*Treponema pallidum* bacteria

#### Symptoms

Chancres, flu-like symptoms, skin rash, fever; if untreated, dementia, heart failure, bone pain

#### Duration

Years, unless treated early

#### Syphilis



*A secondary syphilis rash on the back.* CNRI/PHOTO RESEARCHERS, INC.

Syphilis is a disease that attacks the body in stages:

- Primary syphilis: Acquired by direct sexual contact with an infected person, this stage usually appears after about ten days after the spirochete is transmitted, but can take several months to appear. Its major symptom is the chancre (pronounced SHANK-er), a painless skin ulcer that appears at the location of exposure to the bacterium, usually on the penis, anal area, or vagina. There may be one or several chancres. The patient may also develop swollen lymph nodes, but there are rarely any other symptoms in this stage. Chancres go away after about a week or two even if untreated, but the disease does not.
- Secondary syphilis: This stage develops about six to eight weeks after the primary infection. Its symptoms are the reason syphilis has been called the great pretender—they range from fever, loss of appetite, joint pains, and other flu-like symptoms to a reddish-pink skin rash, hair loss, headache, and swollen lymph nodes. The disease is most contagious in this stage.
- Latent syphilis: Also known as dormant syphilis, in this stage the patient may have no symptoms or occasional flare-ups of the symptoms of secondary syphilis. The patient is not usually infectious but will still test positive for the disease if given a blood test.

## Dr. Hinton's Blood Test

The first modern treatment for syphilis, a drug containing a form of arsenic, was developed in Germany in 1908. Although not as effective as penicillin later proved to be, this useful drug prompted researchers to look for better ways to diagnose syphilis. In the early 1930s, a blood test more accurate than the previously accepted tests was developed by Walter Augustus Hinton (1883–1959), a graduate of Harvard Medical School and the first African American to become a full professor in any department at Harvard. Hinton, the son of two former slaves, acquired an international reputation as a public health expert in the prevention and treatment of syphilis.

Hinton's blood test was not only more accurate, but simpler and less expensive than former tests. It was officially endorsed as the diagnostic standard by the U.S. Public Health Service in 1934. In 1936 Hinton published the first medical school textbook written by an African American; it was on syphilis and its treatment. Hinton continued teaching at Harvard until 1950. After his retirement he worked as a staff physician at a hospital for crippled children in Canton, Massachusetts.

A woman in this stage of syphilis can still transmit syphilis to her unborn child.

• Tertiary syphilis: Between a third and a half of patients with latent syphilis will progress to third-stage, or tertiary, syphilis. This stage can occur as early as one year after the initial infection or as late as fifty years afterward. It is marked by the development of gummas, large noncancerous tissue growths that may occur anywhere on the skin or inside the body, including the skeleton. Untreated tertiary syphilis, which is rarely seen in the 2000s, can lead to movement disorders, heart failure, dementia, and other disorders of the central nervous system. Patients with dementia caused by syphilis usually die within two to three years.

#### Demographics

Syphilis has been on the increase in the United States in recent years. The number of cases dropped after the introduction of penicillin as an effective treatment in the 1940s. In the 1980s, however, the number of cases began to rise again as a result of the growing use of crack cocaine and intravenous drugs, the exchange of

sex for drugs, and the increase in the number of people with multiple sexual partners.

According to the Centers for Disease Control and Prevention (CDC), the number of reported cases of primary and secondary syphilis rose 12 percent between 2005 and 2006. In 2006, there were 36,000 cases of syphilis reported in the United States; most of these occurred in adults between the ages of twenty and thirty-nine. The rates for syphilis were highest in women twenty to twenty-four years of age and in men thirtyfive to thirty-nine years of age. There were 349 cases of congenital syphilis reported in 2006. In the same year, 64 percent of the reported syphilis cases in males were among men who have sex with men (MSM). The rates of syphilis vary in different parts of the United States. The CDC reports that half of all cases of syphilis in the country are found in twenty counties and two cities. Syphilis also disproportionately affects African Americans, who accounted for 41 percent of all cases of the disease in 2006.

Some people are at increased risk of becoming infected with syphilis:

- Men who have sex with men. The highest rates of syphilis in the United States as of 2008 were in this group.
- Those who are HIV-positive. About 50 percent of men diagnosed with syphilis since the early 2000s are also HIV-positive.
- People who drink heavily or use mood-altering drugs. These substances do not cause syphilis, but they do affect a person's ability to think clearly and practice safe sex.
- People who have many different sexual partners or have sex with strangers.

# **Causes and Symptoms**

Syphilis is caused by *Treponema pallidum*, a bacterium that is usually transmitted by direct oral, anal, or vaginal contact with an infected person but can also be transmitted by an infected mother to her baby before birth. The bacteria can cross the mucous tissues lining the mouth and the genitals; they can also enter the body through a crack or break in the skin elsewhere on the body. In addition, the disease can be transmitted through transfusion with infected blood. Syphilis cannot be spread, however, through casual contact with toilet seats, doorknobs, swimming pools, hot tubs, bathtubs, shared clothing, drinking glasses, or food utensils.

As has been noted earlier, the symptoms of syphilis vary depending on the stage of the infection:

- Primary syphilis: one or more chancres, possibly swollen lymph nodes.
- Secondary syphilis: Skin rashes, flu-like symptoms, headache, loss of appetite, fever, fatigue.
- Latent (dormant) syphilis: Flare-ups of symptoms of secondary syphilis or no symptoms at all.
- Tertiary syphilis: Difficulty coordinating muscle movements, paralysis, numbness, gradual blindness, and dementia, possibly ending in death.

#### Diagnosis

Primary syphilis is diagnosed by taking a sample of chancre cells and sending it to a laboratory for examination under a microscope to look for *Treponema pallidum*. This test is also useful in distinguishing the spirochete that causes syphilis from those that cause Lyme disease or yaws, a tropical disease.

Blood tests are more effective in diagnosing secondary than primary syphilis. The two blood tests most commonly used as screeners are called the Venereal Disease Research laboratory Test (VDRL), and the rapid plasma reagin test (RPR). These tests work by showing the presence of antibodies stimulated by the bacteria in the patient's blood. Some other diseases, such as lupus, infectious mononucleosis, and rheumatoid arthritis can give a positive result on these tests, however. For this reason a patient who has a positive test result on either the VDRL or the RPR is usually given another test that is specific for *T. pallidum*.

Patients with tertiary syphilis have their cerebrospinal fluid checked periodically by a spinal tap in order to monitor the progress of treatment for the disease.

#### Treatment

Syphilis can be treated with a single injection of penicillin (or another antibiotic if the patient is allergic to penicillin) during its primary and secondary stages. Patients in the later part of the latency stage or those diagnosed with tertiary syphilis are given three injections of penicillin, each one week apart. Patients with tertiary syphilis that has affected the brain may require intravenous penicillin every four hours for ten to fourteen days.

Some patients, especially with secondary syphilis, have a brief reaction to treatment marked by fever, chills, and headache. This reaction is caused by the death of large numbers of spirochetes in a short period of time. It usually lasts only a day and can be treated at home with aspirin or acetaminophen.

Having syphilis does not make a person immune to it; it is possible to become reinfected with the disease after having been successfully treated for it.

People being treated for syphilis should not have sex with anyone until the doctor confirms that they are no longer infectious, which may take 2–3 months. The usual schedule for follow-up is blood tests at three, six, and twelve months after treatment to make sure that the spirochete has been

# WORDS TO KNOW

**Chancre:** A painless ulcer that forms on the skin during the early stage of syphilis.

**Gumma:** A soft noncancerous growth of tissue found in patients with tertiary syphilis.

Yaws: A spirochete-caused tropical infection of the skin, bones, and joints.

**Congenital:** Present at birth.

eliminated. Patients with tertiary syphilis should have repeat blood tests and tests of spinal fluid every six months for at least three years.

# Prognosis

Antibiotic treatment for primary and secondary syphilis has a very high cure rate. As of 2008, there were no reports of penicillin-resistant strains of *T. pallidum*.

Untreated primary or secondary syphilis has a one in two or one in three chance of progressing to tertiary syphilis and possible death.

# Prevention

Syphilis can be prevented by:

- Abstaining from sexual intercourse, or having sex with only one partner who is uninfected and faithful.
- Using latex condoms when having sex with someone whose health status is not known.
- Avoiding excessive drinking and drug use.

# The Future

Researchers are looking into newer oral antibiotics as effective ways of treating syphilis, particularly in developing countries where injectable penicillin is difficult to obtain or to keep refrigerated. Other areas of research include the development of effective diagnostic tests based on urine or saliva samples rather than blood. Last, the sequencing of the genome (genetic code) of *T. pallidum* in 1998 has encouraged scientists to work on developing a vaccine against syphilis, although such a vaccine lies at least several years in the future.

SEE ALSO AIDS; Chlamydia; Genital herpes; Gonorrhea

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# Τ



Genetic

Infection

Injury

Multiple

. Other

Unknown

#### Also Known As Tendinopathy

#### Cause

Overuse of certain muscles, aging process

#### Symptoms

Pain, warmth, and swelling in affected tendon; pain worsened by activity

**Duration** Days to weeks



# Definition

Tendinitis (also spelled tendonitis) is inflammation or irritation of a tendon—one of the thick cords of white connective tissue that attach muscle to bone. Tendons are less elastic than muscle; they transmit the force of a muscle's expansion or contraction to the bone or joint to cause movement. A tendon becomes inflamed when it is overused or injured by a blow or some other trauma, resulting in warmth, pain, swelling in the affected joint, and some loss of motion in the joint.

# Description

Tendons are located all over the body and range in size from the very small tendons in the hands to large tendons like the Achilles tendon at the back of the heel. The most common locations where tendinitis develops are the shoulders, elbows, knees, wrists, and heels. Some forms of tendinitis are named for the athletic activities that are known to cause them, such as golfer's elbow (inner tendon of the elbow), tennis elbow (outer tendon of the elbow), pitcher's shoulder (rotator cuff tendinitis), and jumper's knee (quadriceps or patellar tendons of the knee). Although these types of tendinitis are associated with sports, they can also develop in people whose occupations require repetitive movements of the hands, arms, or legs. Illustration of tendinitis in the Achilles tendon. © PHOTOTAKE INC. / ALAMY.



Tendinitis begins when microscopically small tears develop in the tissue of the tendon as a result of overuse. In some cases the tendon may have been previously weakened by normal aging or by such diseases as rheumatoid arthritis. If the activity that has caused the first small tears in the tendon is continued, the tendon becomes irritated and inflamed, leading to swelling and pain in the soft tissues surrounding the joint. In most cases, the pain becomes noticeably worse when the joint is moved or used.

# Demographics

Tendinitis is a very common health problem in the general population. No exact statistics are kept because many people treat mild tendinitis at home without going to their doctor. It is known, however, that middle-aged adults are the age group most likely to develop tendinitis.

People who are most at risk of developing tendinitis are:

- Those over forty-five years of age.
- People whose occupations require repetitive movements of the upper arm, wrist, or shoulder. These include carpenters, gardeners, painters, musicians, dancers, manicurists, dental hygienists, and dentists (tennis elbow and golfer's elbow); construction work, electrical repair, or other jobs requiring reaching overhead (pitcher's shoulder); and typing or data entry (wrist tendinitis).

- People with rheumatoid arthritis, osteoarthritis, thyroid disorders, diabetes, or gout.
- People who take a type of prescription antibiotic known as fluoroquinolones.
- People who have been recently injured in an accident or are recovering from burns.
- People who abuse steroids for muscle building.

# **Causes and Symptoms**

Tendinitis is caused primarily by overuse of specific muscles; however, the aging process and diseases associated with joint inflammation may speed up the development of tendinitis or make the symptoms worse.

In general, the symptoms of tendinitis include pain and swelling of the soft tissues near the affected tendon. Symptoms in specific tendons are as follows:

- Golfer's elbow: Pain when the wrist is flexed or turned outward, as when shaking hands or using certain kinds of handheld tools.
- Tennis elbow: Pain spreading into the upper arm or lower arm when the patient grips an object or extends the wrist (as when pouring liquid from a pitcher).
- Achilles tendinitis: Pain in the heel as the foot is flexed while running.
- Tendinitis in the knee: Pain in the side of the knee during downhill running, or pain just below the kneecap during or after physical activity.
- Rotator cuff tendinitis: Pain when the arm is raised above the head; pain worsened when the arm is lowered across the chest.

# Home Care for Tendinitis

There are five points to keep in mind in treating tendinitis at home. The word PRICE is a good way to remember them:

- Protection. Protecting the injured tendon involves the use of slings, splints, crutches, or elastic bandages to prevent unnecessary movement of the affected area.
- Rest. Rest means avoiding activities or body movements that make the pain or swelling worse; it does not mean complete bed rest. Swimming or exercising in a pool is often a good way to keep active without harming the injured tendon.
- Ice. An ice pack can be applied to the affected area for fifteen to twenty minutes every four to six hours for three to five days, or a few days longer if the doctor recommends it.
- Compression. Compression refers to the use of a wrap or Ace bandage to keep swollen muscles from restricting movement in an injured joint.
- Elevation. Raising the affected arm or leg on a pillow or cushion is helpful in relieving swelling in the affected joint.

Although rest is an important part of self-care at home for tendinitis, gentle movement of the affected joint after a few days of complete rest is necessary to prevent stiffness. Doctors recommend moving the sore area slowly and gently through its full range of motion or doing slow stretching exercises.

Nonsteroidal anti-inflammatory drugs, or NSAIDs, can be used to relieve pain and inflammation in the sore joint. These drugs include aspirin as well as Advil and Motrin.

#### Diagnosis

It is important to have tendinitis diagnosed and treated promptly because continued overuse of a damaged tendon can lead to complete rupture (tearing) of the tendon. The doctor will begin by taking a patient history, noting any recent injuries and medications taken as well as any previously diagnosed conditions like diabetes or rheumatoid arthritis.

The next step is a physical examination of the painful joint. This part of the examination involves asking the patient to hold or move the affected limb or joint according to the doctor's instructions while the doctor puts pressure on the muscles and tendons. For example, to detect golfer's elbow, the doctor will ask the patient to place the forearm on the examining table with the inside of the wrist upward. The patient is then asked to make a fist while the doctor holds the forearm down. If the patient has golfer's elbow, he or she will feel pain in the elbow. There are similar tests for the tendons in the shoulder, knee, wrist, and heel.

Imaging tests are not usually necessary unless the doctor wants to check for bone chips or fractures in addition to the tendinitis. If the patient does not get better after rest and medications for the tendinitis, the doctor may order an ultrasound or magnetic resonance imaging (MRI) test. A blood test is usually performed only when the doctor needs to rule out rheumatoid arthritis as the cause of the pain and swelling.

#### Treatment

Treatment of tendinitis is conservative, usually consisting of pain relievers, rest, ice applications, and limiting the movement of the affected joint by splints or a sling (for rotator cuff tendinitis). After recovering from the acute pain, the patient should consider a program of physical therapy and exercise to increase muscle strength and range of motion in the affected joint.

For severe pain that is not helped by NSAIDs, the doctor may inject steroid drugs to decrease pain and inflammation. Steroid injections should not, however, be used more than a few times because they may weaken tendons and increase the risk of a ruptured tendon. Ruptured tendons require prompt surgical treatment to prevent permanent disability.

Other treatments for tendinitis that are currently considered experimental include ultrasound therapy, vitamin E, nitric oxide, and extracorporeal shockwave therapy or ESWT. ESWT was originally developed to treat kidney stones but has been approved by the Food and Drug Administration (FDA) for the treatment of tennis elbow.

#### Prognosis

The prognosis for recovery from tendinitis is very good with appropriate medical treatment and self-care at home.

#### Prevention

The National Institute for Arthritis and Musculoskeletal and Skin Diseases (NIAMS) recommends the following steps to prevent tendinitis:

- Warm up or stretch before exercising.
- Stop and rest if there is pain during a particular exercise or body movement.
- Try different types of exercise; alternate high-impact sports like running or tennis with lower-impact activities like swimming or yoga.
- Make sure that the technique involved in the sport or occupational activity is correct. An improper stance or swing, or poor posture when typing or practicing music, can contribute to tendinitis. If necessary, consult a doctor or physical therapist for guidance.
- Begin slowly when starting a new form of exercise or other physical activity and work up gradually to longer workouts.
- Take frequent breaks from repetitive jobs or chores.
- Cushion joints at risk of tendinitis; use pads for the knees when gardening, padded gloves for heavy tools, and additional grips on golf clubs.
- Use a two-handed grip for heavy tools.
- Do stretching exercises after a workout as well as before, to maintain full range of motion in the joints.

#### The Future

New treatments for the pain of tendinitis are currently being studied. One treatment is an NSAID patch that could be applied to the skin and allow the drug to relieve pain in the affected joint without upsetting the

# WORDS TO KNOW

Achilles tendon: The tendon that connects the calf muscle to the back of the heel. Tendinitis in the Achilles tendon is common in sports that involve running and jumping.

**Tendon:** A thick band or cord of dense white connective tissue that attaches a muscle to a bone.

**Rotator cuff:** A group of four muscles that attach the arm to the shoulder blade.

stomach, which oral NSAIDs often do. Another experimental treatment for tendinitis is injections with tissue plasminogen activator (tPA) factor, an enzyme that is presently used primarily to treat heart disease and stroke by breaking up blood clots.

**SEE ALSO** Diabetes; Rheumatoid arthritis; Sprains and strains; Steroid use

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# Tetanus

# Definition

Tetanus is an infectious disease that, by the production of a potent toxin, affects the muscles of the body, causing stiffness, spasms, difficulty breathing, and a risk of death. It is caused by a bacterium called *Clostri-dium tetani*, which lives in the soil.

### Description

Tetanus has been familiar to doctors for centuries; it was described by Hippocrates around 400 BCE as a common but frequently fatal disease. He was the first medical writer to list the characteristic symptoms of the disease, including the tightening of the jaw muscles that gave tetanus its common name of lockjaw.

Tetanus is caused by an anaerobic bacterium that exists in the soil as a spore and gains entrance to the body through a wound, often a puncture or a crushing wound. There are four basic forms of tetanus: neonatal, which affects newborns; cephalic, which is limited to the head; local, which affects the muscles closest to the infected wound; and generalized, which affects the entire body and can lead to death.

# Demographics

Tetanus was a fairly common disease until the twentieth century. The bacterium that causes tetanus was first identified in 1885, and an effective vaccine was developed in the 1940s. This vaccine helped to make tetanus a rare disease in the United States; only twelve cases of tetanus were diagnosed in American soldiers during World War II, compared to thousands of deaths from tetanus among unimmunized German and Italian troops. The annual number of cases of tetanus among civilians in the United States fell from 600 in 1940 to forty-three in 2000. As of the early 2000s, the rate of tetanus in the United States is 0.16 cases per 1 million population.

Most cases of tetanus in the United States are cases of generalized tetanus; neonatal tetanus is very rare in North America. The adults at greatest risk include intravenous drug users, Hispanics, people over sixty Also Known As Lockjaw

#### Cause

Clostridium tetani bacteria

#### Symptoms

Muscle stiffness and spasms, fever, difficulty breathing, irregular heartbeat, death

Duration Several weeks

#### Tetanus



Baby with muscle stiffness from tetanus in a Vietnamese hospital. SUE FORD/PHOTO RESEARCHERS, INC.

who have allowed their immunization to lapse, and older adults with diabetes.

In the developing world, however, neonatal tetanus is a major killer of newborns, responsible for thousands of deaths. It occurs when the stump of the baby's umbilical cord becomes infected by dirt containing tetanus spores. There are about a million cases of tetanus reported around the world each year, causing between 300,000 and 500,000 deaths. The countries that have the highest rates of tetanus are those with hot, damp climates and soils containing large amounts of animal manure.

#### **Causes and Symptoms**

The bacterium that causes tetanus is anaerobic, which means that it only thrives in the absence of oxygen. It is found in the soil, in dust, in the manure produced by farm animals and household pets, and in the digestive tracts of 10–25 percent of humans. The bacterium forms spores (an inactive stage) that can remain dormant outdoors for years. When the spores get into the body through a puncture wound, an injury that crushes tissue, or a drug injection, they germinate (start to grow) in dead or dying tissue, which lacks oxygen. Germination usually occurs between three days and three weeks after the spores are introduced into the body. The bacteria secret two toxins, one of which affects the nervous system

# The Bridge Builder

John Augustus Roebling (1806–1869) was one of the greatest civil engineers of the nineteenth century, famous for the beauty as well as the strength of the bridges he designed. Roebling left his native Germany in 1831 to seek new opportunities in the United States. Settling near Pittsburgh, Pennsylvania, he began to work for the state as a designer of railroad bridges. He built several suspension bridges over the Allegheny and Monongahela Rivers, using wire ropes and cables that he invented himself.

New bridges were a vital part of efficient troop movement during the Civil War, and Roebling began work on a bridge across the Ohio River at Cincinnati, Ohio, in 1863. It was the longest suspension bridge in the world when completed in 1867. A few months later, Roebling began work on his last project—the Brooklyn Bridge across the East River in New York. The bridge, a worldfamous landmark, cost Roebling his life in 1869. He believed in supervising the construction work himself rather than giving the job to others, and as he stood at the edge of a dock near the construction site a ferry crushed his foot against the dock. Roebling allowed doctors to amputate his crushed toes but refused further medical treatment.



*John Augustus Roebling.* COURTESY OF THE LIBRARY OF CONGRESS.

Twenty-four days later he was dead of tetanus, a disease for which the cause would not be identified for another sixteen years and an effective treatment would not be available for another half century.

and causes the muscle stiffening and spasms that are characteristic of tetanus.

The first symptoms of tetanus usually appear about eight days after infection. The patient typically has a sore throat with difficulty swallowing, headache, stiffness in the jaw muscles known as trismus (lockjaw), followed by stiffness of the neck, heavy sweating, fever, stiffness in the muscles of the abdomen, and spasms in the muscles close to the site of the cut or other injury.

In severe tetanus, the patient has spasms of the arms, legs, and back muscles that can be violent enough to dislocate or break bones. The spasms can also affect the vocal cords and the muscles that control breathing. The patient's blood pressure may alternate between being too high and too low, and the heart rhythm may change from a fast heartbeat to an overly slow heartbeat. Without treatment, the patient may die from respiratory failure or the heart suddenly stopping.

#### Diagnosis

The diagnosis of tetanus is usually based on the patient's history, particularly a recent crushing or penetrating injury, and such symptoms as fever, muscle stiffness, and changes in blood pressure and heart rhythm. The doctor will also check the patient's record of immunization against tetanus.

There are no laboratory tests or imaging studies that are helpful in diagnosing the disease, although the doctor may order a blood test to rule out certain types of poisons.

#### Treatment

Treatment can begin at home with cleaning a wound carefully with soap and running water. A clean cloth can be applied to stop the bleeding. If the wound was caused by a farm tool or other object left outdoors, or resulted from a crushing injury, the injured person should consult their doctor, particularly if they have not had a tetanus booster shot within the past ten years.

Medical treatment begins with prevention, using debridement (surgical removal of any dead or dying tissue that may contain tetanus bacteria). The next step in treatment consists of administering human tetanus immune globulin (TIG), a form of immunization that neutralizes the toxin already produced by the bacteria. TIG produces temporary protection against tetanus in patients who have not been vaccinated against the disease or who have not had a booster shot in the last ten years.

Other drugs that are given to treat severe tetanus include various antibiotics to kill the bacteria directly, muscle relaxants and tranquilizers to prevent muscle spasms, sedatives to help the patient sleep, and painkillers to relieve the pain caused by the muscle spasms.

Patients who are having trouble breathing may be placed on a ventilator and given fluids intravenously to prevent dehydration and provide extra nutrition. The extra calories are needed because the muscle spasms of severe tetanus use up large amounts of the body's energy stores.

# WORDS TO KNOW

**Anaerobic:** Capable of living in the absence of oxygen.

**Debridement:** Surgical removal of dead or dying tissue.

Spore: The dormant stage of a bacterium.

**Trismus:** The medical name for the spasms of the jaw muscles caused by tetanus.

#### Prognosis

The prognosis of tetanus depends partly on the form of the disease. Cephalic and local tetanus have very high recovery rates. Neonatal tetanus, however, has a mortality rate of close to 90 percent. Generalized tetanus causes about five deaths per year in the United States; the overall mortality rate is about 30 percent but is close to 50 percent in people over the age of sixty. If the patient recovers, however, he or she usually recovers completely, without lasting damage to the nervous system.

#### Prevention

Prevention is the most effective defense against tetanus. Children are usually immunized against tetanus by a combination vaccine that also protects against diphtheria and whooping cough. They are usually given five doses of the combination vaccine, at two, four, six, and fifteen months of age, and a final dose at four to six years. They should then have a booster shot every ten years.

Adults who have not had a booster shot within the last ten years should be given a booster shot as well as TIG if they see their doctor about a wound that might be infected by tetanus spores.

#### The Future

Tetanus is unlikely to ever be completely eradicated because the spores that cause the disease are so widely distributed in soils around the world and in the digestive systems of humans and animals. The best hope for lowering the worldwide death toll from this disease is universal immunization and greater availability of appropriate treatments.

**SEE ALSO** Whooping cough

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#### Also Known As

Mediterranean anemia, Cooley's anemia, hemoglobin H disease, hydrops fetalis

#### Cause

Genetic mutations on chromosome 11 or chromosome 16

#### Symptoms

Anemia, childhood jaundice, bone abnormalities, fetal or newborn death

#### Duration

Lifelong

# Thalassemia

# Definition

Thalassemia is the name of a group of inherited blood disorders caused by various mutations in the genes that affect the body's ability to produce hemoglobin, the oxygen-carrying pigment in red blood cells. Some doctors prefer to speak of the thalassemias (in the plural) to underscore the fact that they are a group of blood disorders rather than a single disease.

# Description

There are two major types of thalassemia, categorized according to the part of the hemoglobin molecule that is affected by the genetic

mutations. The hemoglobin molecule has four protein chains, two called alpha chains and two called beta chains. Thalassemias are classified as alpha thalassemias if the genetic mutation damages the alpha chain, and beta thalassemias if the genetic mutation affects the beta chain.

Four genes are needed to make the proteins in the alpha chains of the hemoglobin molecule. If one or two of these genes are defective, the person has what is called thalassemia trait. They will not have the symptoms of thalassemia but will carry the genetic mutation. A child who inherits two thalassemia trait genes (one from each parent) will have the symptoms of the disease. A child of two carriers has a 25 percent chance of receiving two trait genes and developing the disease and a 50 percent chance of being a thalassemia trait carrier.

On the other hand, if the person has defects in more than two of the four genes needed to make the proteins in the alpha chain, they will have moderate to severe anemia. The most severe form of alpha thalassemia is known as alpha thalassemia major or hydrops fetalis. Babies with hydrops fetalis usually die before or shortly after birth.

In beta thalassemia, the person has mutations in one or both of the genes needed to form



the beta chain of the hemoglobin molecule. The severity of beta thalassemia depends on the extent of the changes in one or both genes. If both genes are affected, the result is moderate to severe anemia. The severe form of beta thalassemia is sometimes called thalassemia major or Cooley's anemia. X ray of the hand of a patient with thalassemia. The lighter areas show weakened bones. CNRI / PHOTO RESEARCHERS, INC.

# Demographics

The demographic distribution of alpha thalassemia is different from that of beta thalassemia. The rate of alpha thalassemia in the United States is rising due to immigration from countries where the mutations that cause this type of thalassemia are relatively common. These include countries in Africa, the Middle East, India, Southeast Asia, southern China, and occasionally the countries around the Mediterranean Sea. About 300,000–400,000 severely affected infants are born worldwide every year; more than 95 percent of these births occur in Asia, India, and the Middle East.

Doctors estimate that about 15 percent of African Americans are silent carriers of alpha thalassemia. In addition, thalassemia trait (minor) occurs in 3 percent of African Americans and in 1–15 percent of persons from Mediterranean countries.

Beta thalassemia is more common in Africa, Southeast Asia, Iran, Arabia, Central Asia, and the countries around the Mediterranean than it is in northern Europe or North America. It may affect as many as 10 percent of the population in Southeast Asia and Africa.

Males and females are equally affected by both alpha and beta thalassemia.

#### **Causes and Symptoms**

Both alpha and beta thalassemia are caused by mutations in genes that affect the structure of the hemoglobin molecule. In alpha thalassemia, one or more of four genes (two from each parent) located on chromosome 16 are defective. In beta thalassemia, the mutation is located in the HBB gene on chromosome 11.

The symptoms of alpha thalassemia range from no symptoms in carriers of the thalassemia trait to mild or severe symptoms of the disease, depending on the number of genes that are defective.

- Thalassemia trait. This is a mild form of alpha thalassemia in which the person has red blood cells that are smaller than normal and a mild anemia, but no major health problems. It is often misdiagnosed by doctors as iron deficiency anemia.
- Hemoglobin H disease. This is a form of alpha thalassemia in which the lack of alpha protein is great enough to cause severe anemia and such serious health problems as an enlarged spleen, deformed bones, leg ulcers, gallstones, and fatigue. It is named for the abnormal type of hemoglobin that is formed by the beta proteins in the hemoglobin molecule. Children with hemoglobin H disease are often born with jaundice and anemia; the condition is usually detected shortly after birth.

• Alpha thalassemia major or hydrops fetalis. In this condition there are no functional alpha proteins at all in the patient's hemoglobin. Babies with hydrops fetalis die before or shortly after birth.

Beta thalassemia is classified into three types, thalassemia minor, thalassemia intermedia, and thalassemia major (Cooley's anemia) depending on the severity of symptoms.

- Thalassemia minor: Persons with this form of beta thalassemia usually experience no health problems except for an occasional mild anemia. Their condition is usually discovered only through a routine blood test.
- Thalassemia intermedia: Children with this form of beta thalassemia may require occasional blood transfusions to treat their anemia. Bone deformities and enlargement of the spleen are common.
- Thalassemia major: This is a potentially life-threatening form of beta thalassemia. Children with Cooley's anemia must receive regular blood transfusions and extensive medical care in order to survive past childhood.

# Diagnosis

Both major types of thalassemia are diagnosed by blood tests and genetic testing. Genetic testing can determine whether a person is a carrier of thalassemia even though they may not have symptoms.

Genetic testing can be done before a baby is born, around the eleventh week of pregnancy. The doctor can remove a small amount of tissue from the placenta for analysis. Amniocentesis, which involves taking a sample of the fluid surrounding the baby in the uterus, can be performed around the sixteenth week of pregnancy.

#### Treatment

Treatment depends on the severity of thalassemia. People with trait thalassemia may not have symptoms that need treatment. Patients with hemoglobin H disease and Cooley's anemia typically require frequent blood transfusions for the rest of their lives along with a type of therapy called chelation therapy. Most patients with a major form of thalassemia are given red blood cell transfusions every two to three weeks, as much as fifty-two pints of blood per year. These transfusions are necessary to make sure that the patient's blood is delivering enough oxygen to the tissues to meet energy needs.

Chelation therapy consists of a medication administered to help the body get rid of the iron that builds up in tissues as a result of frequent blood transfusions. If the excess iron is not removed, it will eventually damage the patient's liver and heart and lead to early death from organ failure. Chelation therapy for severe thalassemia involves the use of Desferal, a drug that is infused into the body through a pump worn under the skin of the stomach or legs five to seven times a week for as long as twelve hours. A newer drug for chelation therapy was approved by the Food and Drug Administration (FDA) in 2005. It can be taken by mouth just once a day. Both drugs work by changing the iron into a form that the body can excrete through the urine or stool.

Patients with beta thalassemia major (Cooley's anemia) usually have the spleen removed by surgery after they are six or seven years of age to prevent later complications and lower the frequency of blood transfusions required. Another treatment that is effective if a good donor match can be found is transplantation of bone marrow. Transplantation, however, is possible only for a small minority of patients who have a suitable bone marrow donor. The transplant procedure itself is still risky and can result in the patient's death.

#### Prognosis

The prognosis of thalassemia depends on the type and the severity. Carriers of alpha thalassemia who do not have symptoms have a normal life expectancy and usually enjoy good health, although they should consider genetic counseling before starting a family. Patients with hemoglobin H generally survive into adulthood, although some have a lowered quality of life because of the disease. Hydrops fetalis is almost always fatal.

Patients with mild beta thalassemia have a normal life expectancy with generally good health, although like patients with alpha thalassemia, they should be informed about the hereditary nature of their condition. The prognosis for patients with Cooley's anemia depends on their compliance with frequent transfusions and chelation therapy. Some people find Desferal therapy so troublesome and painful that they give up on treatment. Untreated, Cooley's anemia usually leads to death from heart failure or infection before age twenty.

### WORDS TO KNOW

- **Anemia:** A condition in which there are not enough red cells in the blood or enough hemoglobin in the red blood cells.
- **Chelation therapy:** A form of treatment to reduce overly high levels of iron (or other metals) in the body by giving the patient a chemical that allows the body to get rid of the excess metal in urine or stool.

**Cooley's anemia:** Another name for the most severe form of beta-thalassemia.

- **Hemoglobin:** A pigment in red blood cells that contains iron and transports oxygen from the lungs to other body tissues.
- **Hydrops fetalis:** The most severe form of alpha thalassemia, leading to death before or shortly after birth.
- **Thalassemia trait:** A condition in which a person is missing one or two genes required to make the proteins in the alpha chains of the hemoglobin molecule. The person does not have the symptoms of thalassemia but can pass the genetic deficiency to their children.

#### Prevention

There is no way to prevent either alpha or beta thalassemia.

#### The Future

Researchers are hoping to develop a form of gene therapy to treat patients with beta thalassemia. This type of treatment would involve inserting normal HBB genes into immature bone marrow cells. These are the cells that produce red blood cells; thus gene substitution would lead to the eventual production of normal red blood cells.

#### **SEE ALSO** Anemias

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# Tick-borne Diseases

Tick-borne diseases are infectious diseases carried by small eight-legged insects belonging to the spider family. Ticks are parasites that feed on blood and usually live in weeds, shrubs, or tall grass. They cannot fly and must attach themselves directly to a deer, human, dog, or other animal passing through the grassy area. Ticks have a harpoon-like structure in their mouths that enables them to attach to their host and feed for several hours or even days.

Tick-borne diseases include those caused by bacteria (Lyme disease), viruses (various forms of encephalitis), and rickettsiae (Rocky Mountain spotted fever), which are a type of bacterium. These diseases can be difficult to diagnose because a tick may carry more than one disease organism, thus infecting a human with two diseases at the same time. Most tick-borne diseases are treated with antibiotics before the specific disease organism is identified because delayed treatment can lead to such complications as arthritis and severe fatigue. Rocky Mountain spotted fever is potentially fatal.

Tick-borne diseases are becoming increasingly common in the United States as people build homes and move into areas that were recently wilderness. Controlling the deer population, wearing protective clothing, and clearing brush and tall grass where possible are important preventive strategies.

**SEE ALSO** Encephalitis; Lyme disease



# Tonsillitis

# Definition

Tonsillitis is an inflammation of the tonsils at the back of the throat caused by infection by a virus or bacterium. The tonsils themselves are two areas of lymphoid tissue; they are part of the lymphatic system and act as part of the immune system. Tonsillitis may be caused by either viruses (about 75 percent of cases) or bacteria (the remaining 25 percent).

Recurrent tonsillitis is defined as seven episodes of tonsillitis in one year; five episodes in two successive years; or three infections per year for three straight years.

### Description

The tonsils are part of the immune system protecting the body against infections of the throat and upper respiratory tract. These areas of tissue normally function to neutralize disease organisms before they reach the lower throat; however, they can be overwhelmed by a virus or bacterium. At that point the tonsils become red and swollen, and may develop abscesses, which are pus-filled pockets.

Whether caused by viruses or bacteria, all forms of tonsillitis are contagious. Tonsillitis usually spreads from person to person by contact with discharges from the nose or throat of an infected child or adult. It is often spread by carriers—children or adults who carry a disease agent but do not have any of the symptoms of the disease.

# **Demographics**

Tonsillitis is a common illness among children in the United States; nearly all have at least one episode of tonsillitis by the time they reach the teen years, although tonsillitis is rare in children younger than two. Recurrent tonsillitis is less common, occurring in 11–13 percent of children.

Tonsillitis caused by bacteria is most common in children between the ages of five and fifteen, while children younger than five are more likely to have viral tonsillitis. As far as is known, tonsillitis affects boys Also Known As Tonsillar sore throat, infection of the tonsils

#### Cause

Infection by viruses or by streptococcal bacteria

#### Symptoms

Sore throat, painful swallowing, fever, "hot potato" voice, sore lymph nodes in neck

Duration

Three to twelve days

Enlarged, reddened tonsils due to tonsillitus. © MEDICAL-ON-LINE / ALAMY.



and girls equally, and is equally common in all races and ethnic groups in the United States.

Children at increased risk of tonsillitis include those with malnutrition or weakened immune systems as well as those who attend schools or day care centers with other children who may be carriers.

#### **Causes and Symptoms**

About three-quarters of all cases of tonsillitis are caused by viruses, with the remaining quarter caused by bacteria. The most common virus that causes tonsillitis is the Epstein-Barr virus (EBV), often as part of infectious mononucleosis. Viral tonsillitis can also be caused by the measles virus, herpes virus (the virus that causes cold sores), or adenoviruses (viruses that also cause stomach flu). The most common organisms responsible for bacterial tonsillitis are group A streptococci, the bacteria that cause strep throat and scarlet fever.

The symptoms of tonsillitis may include:

- Sore throat
- Pain in the ears
- Fever and chills
- Red, swollen tonsils
- White or yellow patches on the tonsils

- Pain or difficulty in swallowing
- Bad breath
- Swollen lymph nodes in the neck
- Tiredness and overall sick feeling
- Difficulty breathing or disturbed sleep, if the tonsils are extremely swollen

### Diagnosis

The diagnosis of tonsillitis is made in the doctor's office by a physical examination of the patient's mouth and throat followed by a rapid strep test or culture to identify the disease organism that is causing the infection. During the physical examination, the doctor will feel the lymph nodes in the patient's neck as well as taking the temperature and looking into the throat with the help of a tongue depressor. Redness and swelling of the tonsils, along with pus or other discharges, will be visible to the doctor without the need for special equipment.

To see whether the infection is caused by strep throat bacteria, the doctor will take a throat swab and send the sample of the patient's throat secretions to a laboratory for culture. The doctor may also perform a rapid strep test in the office. This test is faster than a throat culture but is slightly less accurate.

The doctor may also order a blood test to check for signs of a viral infection. An unusually high number of certain white blood cells called

# Did You Know?

Tonsillectomy, or removal of the tonsils, is one of the oldest surgical procedures in Western medicine. The earliest description we have of a tonsillectomy was written between 30 and 50 A.D. by Celsus, a Roman doctor, who invented a hook for grasping tonsils. After hooking the infected tonsil, Celsus removed it with a sharp knife or scalpel. His patient was then given a mixture of vinegar and herbs to stop the bleeding, cleanse the throat, and lower the risk of infection. By the seventeenth century, European doctors had invented an instrument called the tonsillotome, which allowed the doctor to grasp the tonsil and remove it in one motion. The doctor needed an assistant to hold the patient still, however, as anesthesia would not be developed for another two centuries.

Surgeons in the United States generally remove tonsils with forceps and scissors after the patient has been put under general anesthesia. Several new techniques for tonsillectomy have been developed, however, that include the use of ultrasonic scalpels, lasers, and radiofrequency ablation. Radiofrequency ablation is a method that uses radiofrequency energy to shrink or destroy the tissue of the tonsils without the need for cutting.

atypical lymphocytes in the sample may indicate that the tonsillitis is caused by the mononucleosis (Epstein-Barr) virus.

### Treatment

It is important for the doctor to know whether the tonsillitis is caused by strep throat bacteria or viruses because the treatments are different. If the child has viral tonsillitis, treatment consists largely of self-care at home: drinking plenty of warm fluids, taking nonaspirin pain relievers (Tylenol or Advil) to bring down the fever, and gargling with salt water to relieve the sore throat. It may take a week or so for the child to feel better. Antibiotics do not help in treating viral infections.

If the doctor determines that the tonsillitis is caused by a bacterium (almost always the group A streptococcus), he or she will prescribe a course of antibiotics, usually for ten days. If the child has trouble swallowing, the antibiotic may be given by injection, or the doctor may also prescribe a steroid medication to bring down the throat swelling. The child should not return to school or day care for twenty-four to fortyeight hours after beginning antibiotic treatment, to prevent spreading the infection to others. In addition, it is important for the child to take the full course of antibiotics even though he or she may feel better in a day or two.

The doctor may recommend tonsillectomy (surgical removal of the tonsils) for patients with recurrent tonsillitis. Tonsillectomies are not performed nearly as often as they were in the 1950s and 1960s; the number of tonsillectomies performed each year in the United States has dropped from several million in the 1970s to approximately 600,000 in the late 1990s. Tonsillectomies are still recommended, however, to treat severe cases of recurrent tonsillitis accompanied by airway obstruction, and to reduce the risk of ear infections and other potential complications of tonsillitis.

#### Prognosis

Most children and teenagers with tonsillitis recover without any longterm problems. In a few cases, however, the infection can spread from the tonsils to the deeper tissues of the neck or chest. If the tonsillitis is caused by streptococci, it can lead to such complications as inflammation of the kidney or rheumatic fever. For this reason, children with a sore throat and other symptoms of tonsillitis should see the doctor at once.

Tonsillectomy is the second most common operation performed on school-age children and is an extremely safe procedure. It is often done as an outpatient procedure; however, the child may need to recover at home for a week or two afterward.

#### Prevention

People can lower their risk of tonsillitis by washing their hands frequently, avoiding sharing drinking glasses and food utensils, covering the

# WORDS TO KNOW

**Group A streptococcus:** A sphere-shaped bacterium that grows in long chains and causes strep throat as well as scarlet fever and some forms of tonsillitis. Tonsillectomy: Surgical removal of the tonsils.

nose and mouth when coughing or sneezing, and avoiding close contact with others who have upper respiratory infections.

#### The Future

A major concern among doctors is the overuse of antibiotics in treating tonsillitis and other throat infections. Across the United States, 70 percent of children with sore throats who are seen by a doctor are treated with antibiotics even though 30 percent at most have bacterial infections. It can be difficult to distinguish between bacterial and viral infections of the throat even when a throat culture is performed because some people may be carriers of group A streptococci and have the bacteria in their throat at the same time that they have tonsillitis caused by a virus. The chief danger from the overuse of antibiotics is the creation and spread of drug-resistant disease organisms.

**SEE ALSO** Infectious mononucleosis; Scarlet fever; Sore throat; Strep throat

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# Tooth Decay

# Definition

Tooth decay, also known as dental caries, is a disorder of the mouth in which bacteria living in the mouth break down carbohydrates in food. The bacteria produce acids that eat away at the enamel—the hard smooth outer surface of the tooth—eventually producing cavities in the enamel. If untreated, cavities can lead to toothache, loss of the tooth, and bacterial infections spreading to the gums and other tissues.

#### Description

Tooth decay begins with plaque, a thin film that is a mixture of saliva, bacteria, and dead cells from the tissues that line the mouth. When a person eats foods rich in carbohydrates—sugary or starchy foods—the bacteria in the plaque ferment the sugars in these foods, forming lactic acid. Plaque builds up on teeth if it is not removed by regular flossing and brushing, eventually hardening into a whitish substance called tartar.

The surfaces of human teeth are basically composed of minerals that are sensitive to acid. Under normal circumstances, minerals lost from the tooth surface are replaced by the saliva. But when plaque is not removed from the teeth by brushing or flossing, the acid formed by the bacteria begins to remove enamel from the surface of the tooth faster than the

# Also Known As

Dental caries, tooth cavities

#### Cause

Acids produced by bacteria living in the mouth

#### Symptoms

None (in early stages); pain on eating or chewing, tooth sensitivity, toothache (later stages)

#### Duration

Months to years, or until treated

#### Tooth Decay



*A wisdom tooth showing decay.* © DAVE JEPSON / ALAMY.

saliva can restore it. At this point the surface of the tooth begins to develop small pits or cavities. If these are not attended to, they can grow larger and move inward to affect the dentin, a layer of hard tissue below the enamel. Dentin is softer than enamel and protects the pulp at the center of the tooth. When the cavity cuts through the enamel into the dentin, it speeds up the decay process. The pulp contains nerve endings that become inflamed when the bacteria from the mouth are able to gain entry.

Once the tooth decay reaches the pulp, the person may experience pain when biting down on food or the more severe toothache. If the inflammation has spread to affect the jawbone underlying the tooth, the person may develop a pus-filled hollow known as an abscess. In extreme cases, the bacteria may spread to other parts of the face or enter the bloodstream and spread infection to other parts of the body.

#### **Demographics**

Tooth decay is one of the most common health problems around the world. It is also one of the oldest. Prehistoric humans appear to have had dental problems dating from the time that they first began to cultivate grains for food, since bread and flour are rich in carbohydrates. Skulls found in Asia dating from 7000 BCE have teeth that contain holes created by early dental drills. Ancient Egyptian, Indian, and Sumerian medical

# The Dentist Who Discovered the Cause of Tooth Decay

The bacterium that causes tooth decay was identified in the early 1920s by military dentist Fernando Rodríguez Vargas (188–1932). Born in Puerto Rico, Rodríguez Vargas graduated from the dental school of Georgetown University in Washington, D.C. in 1913. He began to investigate the cause of tooth decay in 1915 while working for the federal government's Indian Medical Service in Tucson, Arizona. There he noticed that the Native Americans he was treating had badly discolored teeth as well as a high number of cavities. His early research was interrupted when the United States entered World War I in 1917, and he was sent overseas to examine and treat American soldiers for dental problems.

In 1921, Rodríguez Vargas was working as a bacteriologist in Washington, D.C., for the Army Dental Corps when he discovered that three species of Lactobacillus bacterium are responsible for producing the acids that cause tooth decay. He published his findings in a military medical journal. In 1928 he wrote an article for the *Journal of the American Medical Association* on the usefulness of various antiseptics in cleansing the mouth and preventing tooth decay.

Rodríguez Vargas died of pneumonia in 1932. He has been honored for his pioneering work in dentistry by the Walter Reed Army Institute of Research and the Puerto Rico College of Dental Surgeons. texts attributed tooth decay and cavities to a "tooth worm." The rate of tooth decay among humans increased rapidly after 1850 *CE*, when people began to eat larger quantities of refined sugar, refined flour, and other sweet and sticky foods.

According to the Centers for Disease Control and Prevention (CDC), tooth decay is the most common chronic health disorder among children in the United States between the ages of five and seventeen. Fifty-nine percent of children in this age group have one or more cavities. Many adults also have untreated tooth decay—27 percent of those between the ages of thirty-five and fortyfour, and 30 percent of those over sixty-five.

Worldwide, about 90 percent of schoolchildren and 95 percent of adults have had at least one dental cavity. The rates are highest in Asia and Latin America and lowest in Africa. Males and females are equally likely to develop tooth decay.

In developed countries, the rates of dental cavities have dropped since the 1950s; this decrease is attributed to improved patient education about care of the mouth and teeth, and to such preventive practices as adding fluoride to the water supply. On the other hand, however, the reduction in the number of cavities is not equally distributed throughout the population. Studies in Western Europe, Canada, and the United States indicate that 20 percent of the population in these countries has 70 percent of the cavities.

People at increased risk of tooth decay include:

• Babies who are given sweetened juices or other liquids to drink. "Baby bottle tooth decay" is a common pattern of cavities in the front of the mouth found in very young children.

- People who abuse methamphetamine, an addictive stimulant drug. Methamphetamine dries out the tissues of the mouth, contributing to outright tooth loss as well as tooth decay.
- People with diabetes or Sjögren syndrome. These disorders reduce the amount of saliva in the mouth.
- People with eating disorders. People with bulimia who force themselves to vomit repeatedly weaken the enamel on their teeth by exposing the tooth surfaces to stomach acids.
- Smokers. Smoking causes the gums to recede, thus exposing more of the tooth surface to bacteria in the mouth and the acid they produce.

# **Causes and Symptoms**

Tooth decay is caused by acid-forming bacteria in the mouth that live in plaque that has not been removed from the surfaces of the teeth. Decay is more likely to affect the teeth in the back of the mouth (the molars and premolars). These teeth have pits and grooves on their upper surfaces that make it more difficult to remove plaque completely.

Tooth decay can develop over a period of months or even years without any obvious symptoms. When the decay begins to affect the dentin or the pulp beneath the enamel, however, the patient may begin to notice such symptoms as:

- Sensitivity when eating or drinking sweet, very hot, or very cold foods
- Pain when biting on something firm or tough
- Pus around a tooth, which indicates that at abscess has formed
- Pain that lasts after the person has finished eating
- Visible pits or holes in the tooth
- Bad breath or a bad taste in the mouth
- Intense toothache

# Diagnosis

The diagnosis of tooth decay is made by a dentist rather than a primary care doctor in most cases. The dentist will ask the patient about tooth sensitivity or pain. In some cases he or she can see signs of tooth decay just by looking inside the patient's mouth with an angled mirror. Another tool that is used is

a hook-shaped instrument called an explorer, which allows the dentist to probe soft spots or areas of discoloration in the teeth. Last, the dentist will take x-ray films of the patient's mouth to get a clearer picture of whether and how far the areas of decay have penetrated the tooth.

#### Treatment

The treatment of tooth decay depends on the extent and severity of the problem. If the decay has just begun, the dentist may apply a solution of fluoride to stop the decay process. If the decay has progressed further, the dentist may need to drill away the decayed material or even remove the tooth.

A small area of tooth decay is filled with a silver alloy, a composite resin, gold, or porcelain. Dentists call a filling a dental restoration. If much of the tooth must be removed, the dentist will drill away the top of the tooth and replace it with what is called a crown or a cap. If the tooth decay has affected the pulp in the center of the tooth and destroyed the nerve endings, the dentist removes the pulp along with any decayed portions of the tooth and fills the center of the tooth with a sealing material. This procedure is called a root canal.

If the tooth is badly decayed, was broken in an accident, or is likely to cause trouble in the future, the dentist will perform an extraction. The tooth is lifted with a tool called an elevator and removed from its socket with dental forceps. The dentist will apply a material called gelfoam to speed clotting, or close up the socket with sutures if the wound is large. The patient is given antibiotics to take for several days to prevent infection.

#### Prognosis

The prognosis of tooth decay depends on the stage at which it is discovered and treated. Dental fillings usually last for years with proper care, although they often need replacement when the patient is middle-aged. It is rare for tooth decay to lead to serious complications other than the loss of teeth; however, in a few cases bacteria from a tooth abscess can spread to the tissues of the floor of the mouth or to a hollow space within the brain that lies behind the upper jaw. These infections are potentially life-threatening.

#### Prevention

Tooth decay is one of the most easily prevented health problems. There are several known ways to lower the risk of dental cavities:

# WORDS TO KNOW

- **Abscess:** A collection of pus that has formed in a body cavity or hollow.
- Caries: The medical name for tooth cavities.
- **Dentin:** A firm tissue that lies between the enamel and the pulp of a tooth.

**Enamel:** The hard, smooth, white outer surface of a tooth.

- **Plaque:** A film that forms on the surface of teeth containing bacteria, saliva, and dead cells.
- **Pulp:** The soft living material in the center of a tooth that contains blood vessels and nerve endings.

Tartar: Hardened plaque.

- Brushing the teeth after each meal or snack and using dental floss once a day. Brushing is important because plaque starts to form within twenty minutes of finishing a meal. Flossing helps to remove food particles trapped between teeth.
- Limiting sweets and sugary drinks like soda or sweetened tea.
- Quitting smoking.
- Using a toothpaste that contains fluoride or having the dentist apply a fluoride solution to the teeth during a checkup. Fluoride is a chemical that helps to prevent tooth decay by protecting the minerals in tooth enamel. According to the CDC, people in communities that have added fluoride to their drinking water have 29 percent fewer cavities.
- Sealants. Sealants are protective plastic coatings applied to the surfaces of the back teeth that are most likely to develop cavities. They need to be replaced every few years.
- Antibacterial mouthwashes. Dentists sometimes recommend these for people who are vulnerable to tooth decay because of their medical conditions.
- Having regular dental checkups and necessary treatments.

#### The Future

Newer methods for the prevention of tooth decay include the use of argon lasers to remove early signs of the erosion of tooth enamel and the development of a vaccine against tooth decay. Clinical trials for such a vaccine started in May 2006, but the vaccine has not yet been approved for use.

SEE ALSO Bulimia; Periodontal disease; Sjögren syndrome

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# U

# **Tourette Syndrome**

# Definition

Tourette syndrome (TS) is a disorder of the nervous system characterized by tics, which are involuntary repetitive movements or sounds. It is named for a French neurologist, Georges Gilles de la Tourette (1859– 1904), who first described it in a group of nine patients in 1885.

#### Description

Tourette syndrome is a disorder of the nervous system known for the motor and vocal tics of patients diagnosed with the syndrome. It can affect adults as well as children, but is most commonly diagnosed in children around age seven or slightly older. In most cases TS improves as the child matures and disappears completely by adulthood. Tourette syndrome is considered a spectrum disorder because the symptoms vary in severity from patient to patient; in fact, some doctors think that the diagnosis of Tourette syndrome is often missed because the patient's symptoms are very mild and not noticed by other people. For example, parents often attribute such tics as eye blinking or sniffing noises to vision problems, colds, or allergies.

The tics that characterize TS are sudden, rapid, repeated movements or voice sounds that the person cannot control. The most common first tic in children is a facial tic like eye blinking or grimacing. Motor (movement) tics usually occur before vocal tics, and usually affect the face and neck before moving downward to the arms, chest, or legs. Other common tics include throat clearing, grunting, barking, sniffing, tongue clicking, arm thrusting, shoulder shrugging, jumping, twirling, or kicking. Contrary to popular depictions of TS in the media, the repetition of dirty words, ethnic insults, or curses—known as coprolalia—occurs in only a small minority (about 15 percent) of patients with TS.

The tics may increase or decrease in both number and severity, often disappearing for weeks and months and returning when the person is under stress. Although tics are involuntary, some patients are able to suppress them or manage them for brief periods of time. These patients often feel tension building up when they try to control their tics, however, and will often say that they feel they must release the tension at some point and express the tic. Tics do not go away completely when the patient is sleeping but are usually much less noticeable during sleep.

#### Demographics

At one time TS was thought to be a rare condition but is now diagnosed in 1–2 percent of children and adults in the United States. According to the National Institute of Neurological Disorders and Stroke (NINDS), about 200,000 Americans have the most severe form of TS. Some doctors think that as many as 10 percent of Americans may have mild TS. Also Known As TS, Gilles de la Tourette syndrome

Cause Possibly genetic

Symptoms Motor and vocal tics

**Duration** Years

#### **Tourette Syndrome**

These boys both have Tourette syndrome, but in most ways are typical teenagers. AP IMAGES.



Babies born prematurely appear to be at increased risk of developing Tourette syndrome in childhood, although the reasons for the connection are not yet clear.

TS appears to be equally common in all racial and ethnic groups; however, boys are four to ten times more likely to develop TS than are girls. The reasons for the gender difference are not known.

#### **Causes and Symptoms**

The cause of Tourette syndrome is not known. At one time, the disorder was thought to be a psychiatric illness, but most researchers now believe that TS results from a combination of genetic factors and differences in brain functioning. It is known that the disorder runs in families; however, no specific genes associated with the syndrome have been identified. Brain imaging studies suggest that children with TS have brains that are unusually sensitive to a neurotransmitter (brain chemical) called dopamine, and that the chemical activates parts of the brain that are responsible for the movements involved in tics. One finding that supports this theory is that medications that block the brain's uptake of dopamine are helpful in controlling tics.

Tourette syndrome often coexists with other neurological disorders like attention-deficit hyperactivity disorder (ADHD) and obsessive-compulsive disorder (OCD). About 40 percent of children diagnosed with TS have only TS; the remaining 60 percent have either TS plus ADHD, or TS plus OCD. A few children with Tourette syndrome also suffer from disorders of impulse control, which may include aggressiveness, sexual acting out, or rage attacks.

The tics characteristic of TS are sometimes categorized as simple or complex tics as well as motor or vocal tics. A simple tic is sudden, brief, and involves only a few muscles. A complex tic is a pattern of movement that involves several groups of muscles and takes slightly longer to perform than a simple tic. Examples of simple tics would be eye blinking (motor) or barking (vocal); examples of complex tics would be flapping the arms (motor) and repeating someone else's words (vocal).

#### Diagnosis

There are no laboratory or imaging tests that can be used to diagnose Tourette syndrome, although the patient's doctor may order an ECG or a blood test to rule out seizure disorders or thyroid problems. The doctor will base the diagnosis of Tourette syndrome on a combination of the patient's symptom history, a history of other family members with TS, and the patient's age. The diagnostic criteria used by the American Psychiatric Association specify that the patient must:

- Have several motor tics and at least one vocal tic for a period of a year, with no more than three consecutive months without a tic.
- Have been under eighteen years of age when the tics started.

• Not be abusing alcohol or drugs, or have any other disease or disorder that could be causing the tics.

It is not unusual for people with mild TS to go undiagnosed for years, read about the syndrome and recognize that they have tics, and visit their doctor to have the diagnosis confirmed.

### Treatment

Mild TS may not need any treatment. Although there are medications that can be prescribed to control severe tics, most doctors prefer to wait for a few months rather than prescribe medications right away. One reason is that the drugs given to control tics all have side effects, particularly weight gain, drowsiness, and difficulty concentrating in school. Another reason is that many children with TS have periods when the tics are less severe.

Many doctors recommend psychotherapy for children with TS to help them cope with the social embarrassment and difficulties with schoolwork that they may be facing. Although TS does not cause depression or learning problems, children may become depressed because of rejection by classmates due to the tics, or they may have trouble learning their lessons in school because their attention is focused on controlling the tics.

#### Prognosis

Tourette syndrome is not usually a disabling condition, although it can cause social embarrassment to affected children and teenagers. TS is often most severe in the patient's early teens, with symptoms improving in the later teens and improvement continuing into adult life. About a third of adults find that their tics eventually go away; about 10 percent, however, do not experience significant improvement in their thirties or even in their forties.

People with Tourette syndrome have a normal life expectancy. The disorder does not affect a person's basic intelligence or prevent them from completing their education once they are diagnosed; in fact, many people with Tourette syndrome have above-average intelligence. One follow-up study of thirty-one adults with TS found that all the patients finished high school; 52 percent finished at least two years of college, and 71 percent were employed full time or were pursuing advanced degrees. The chief problem confronting people with TS in adult life is an

# WORDS TO KNOW

**Coprolalia:** The medical term for uncontrollable cursing or use of dirty words.

Involuntary: Not under the control of the will.

**Neurotransmitters:** Chemicals produced by the body that transmit nerve impulses across the gaps between nerve cells.

increased risk of depression, mood swings, and panic attacks compared to the general population.

#### Prevention

There is no known way to prevent Tourette syndrome because its causes are not yet fully understood.

### **The Future**

A newer form of treatment for TS that is considered experimental as of 2008 is deep brain stimulation or DBS. In DBS, a battery-operated device is implanted in the brain to deliver carefully targeted electrical stimulation to the parts of the brain that control movement. In addition to clinical trials of DBS and some newer drugs as treatments for Tourette syndrome, other researchers are using advanced imaging techniques to study the parts of the brain involved in tics. Still other scientists are looking for specific genes that may be linked to TS.

**SEE ALSO** Attention-deficit hyperactivity disorder; Obsessive-compulsive disorder; Prematurity

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**Tic:** A sudden repetitive movement or utterance. Tourette syndrome is considered a tic disorder.

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# **Toxic Shock Syndrome**

#### Definition

Toxic shock syndrome, or TSS, is a life-threatening illness caused by a bacterium called *Staphylococcus aureus*. A similar illness called toxic shock-like syndrome, or TSLS, is caused by another bacterium called *Streptococcus pyogenes*. Both illnesses are forms of sepsis, an inflammatory response of the entire body to severe infection.

#### Description

Toxic shock syndrome, or TSS, was first given its present name by a pediatrician in Denver, Colorado, in 1978. He had been studying a case series of a severe illness caused by *Staphylococcus aureus* in a group of three boys and four girls between the ages of eight and seventeen. The doctor had been able to identify the bacterium in the tissues lining the noses, throats, or vaginas of the patients but not in their blood or urine samples. This finding led him to suspect that the symptoms of the illness—sudden high fever, shock, and a reddish rash resembling sunburn—were caused by a toxin produced by the bacteria. The doctor did not, however, suspect a connection between the illness and menstruation even though three of the four girls in his series had been using tampons at the time they fell ill.

Also Known As TSS

#### Cause

Toxin produced by *Staphylococcus aureus* bacteria

#### Symptoms

High fever, low blood pressure, sunburn-like skin rash, seizures, multiple organ failure

#### Duration

One to two weeks for initial episode; recurrences may occur four to six months later The link between TSS in women and menstruation became clear in late 1979 and early 1980 following the introduction of a new type of tampon made from a superabsorbent type of cotton fiber combined with compressed beads of polyester. The new product was able to absorb 20 times its weight in fluid. By January 1980, doctors in Wisconsin and Minnesota were reporting cases of toxic shock syndrome in women using the new tampons, and doctors in other parts of the United States were reporting



cases of TSS in women using other brands of highly absorbent tampons. The Centers for Disease Control and Prevention (CDC) began to investigate the reports and had the new tampon withdrawn from the market. By September 1980 the number of cases of TSS began to decline as women began to use sanitary napkins in place of tampons. According to CDC statistics, 942 women were diagnosed with tampon-related TSS in the United States from March 1980 to March 1981; forty of the women died.

It is now known that TSS can develop in males and in women who are not menstruating if they become infected with *S. aureus* following surgery or injury to the nasal passages. Patients with TSS develop a sudden high fever, a skin rash that looks like sunburn, a drop in blood pressure, and failure of multiple organ systems in the body.

Patients with toxic shock-like syndrome, caused by *S. pyogenes*, the same organism that causes strep throat, have similar symptoms. They are more likely to fall sick following a skin infection caused by *S. pyogenes*.

# Demographics

TSS related to menstruation is much less common in the United States than it was in the early 1980s. According to the Centers for Disease Control and Prevention (CDC), there were only five documented cases of toxic shock syndrome related to menstruation in 1997, compared to 814 in 1980. As of the early 2000s, the CDC estimates that between one and seventeen of every 100,000 menstruating women will get TSS.

Some people are at increased risk of TSS:

- Women who are menstruating
- Women who use diaphragms or other barrier methods of birth control

*Skin peeling on the hands from toxic shock syndrome.* © MEDICAL-ON-LINE / ALAMY.

# A Survivor's Story

A Canadian woman thought that frequent changes of the high-absorbency tampons she used would be an adequate safeguard against toxic shock syndrome (TSS). However, the first Saturday in November 2004, she fell ill, stating, "[F]or some odd reason I knew it was TSS. I was vomiting all the time and knew it was more than a stomach bug. . . . My husband was taking care of me, following the doctor's instructions to keep me hydrated; give me nausea medication; and not to treat the diarrhea.... I don't remember much of that night. except extreme muscle pain, high fever and my mouth was so sore that I couldn't move my tongue without extreme pain. Every time I sat up I would black out. The next seven days were a blur of pokes, prods and doctors shaking their heads in confusion."

She continued, "After a week in intensive care and recovery I was well enough to go home. Even though the skin was peeling off my palms and the soles of my feet (typical symptoms of TSS), the doctors still were 'not sure' what I'd had!" Her recovery was slow: she gained 40 pounds due to the fluid treatment, had decreased energy, weakness in her hands, extreme pain in her arms and wrists, her hair fell out in handfuls, and she needed antidepressants. In an update from 2006, her hair had finally grown back after a year of complete baldness, she was back to her normal weight, and she no longer needed antidepressants.

- People who are having surgery on the nose
- People with diabetes
- People with a weakened immune system
- People who have developed a staphylococcal infection following surgery

### **Causes and Symptoms**

The cause of toxic shock syndrome is infection with *S. aureus*, while TSLS is caused by *S. pyogenes*. In both illnesses, the bacteria produce exotoxins—toxic substances released into the bloodstream—that overstimulate the body's immune system. As the immune system overreacts to the exotoxins, the person goes into shock, a medical emergency in which the circulatory system starts to shut down and deprives the body's tissues of blood. Without the oxygen and nutrients carried by the blood, the body's organs and tissues begin to fail.

The symptoms of toxic shock syndrome include:

- Sudden high fever of 102°F (38.8°C) or higher
- Nausea and vomiting
- Low blood pressure
- Confusion and disorientation
- Sunburn-like rash that leads to peeling of the skin on the hands and feet after a week or so
- Sore, aching muscles
- Lightheadedness or dizziness when standing up
- Cramping or pain in the abdomen
- Redness in the eyes, mouth, and throat
- Headaches
- Seizures

- Large amounts of watery diarrhea
- Shock occurring about two days after the fever and other symptoms begin

The symptoms of TSLS are similar to those of toxic shock syndrome. In addition, patients with TSLS may have severe pain at the location of the skin infection that precedes TSLS.

# Diagnosis

There is no single test that is used to diagnose TSS or TSLS. The doctor will base the diagnosis on the patient's recent history and a set of criteria drawn up by the CDC to distinguish toxic shock syndrome from measles, Rocky Mountain spotted fever, or other similar diseases:

- Fever of 102.2°F (39°C) or higher.
- Systolic blood pressure below 90 mmHg (Systolic is the top number in a blood pressure reading.)
- Reddish rash followed by skin peeling
- Involvement of three or more of the following organs or organ systems: digestive tract; kidneys; liver; soft tissues lining the throat, nasal passages, or vagina; central nervous system; or the blood

# Treatment

TSS and TSLS are medical emergencies. The patient is taken at once to the hospital emergency department and tampons, nasal packing, or dressings covering a skin infection are removed. The doctor will usually order a blood test to evaluate liver function, kidney function, and the number of white blood cells present. A high white blood cell count, a high level of liver enzymes, and signs of abnormal kidney function suggest a diagnosis of TSS. The doctor will also check the patient's blood pressure, temperature, and heart rate. A female patient will usually be given a pelvic examination.

Treatment begins with locating the source of the infection (vagina, nose, throat, skin) and administering intravenous antibiotics. Depending on the patient's condition, he or she may be placed in an intensive care unit and given one or more of the following:

- Intravenous fluids to raise blood pressure and prevent dehydration
- Monitoring of kidney and liver function

- Dialysis in case of kidney failure
- Oxygen in case of breathing difficulties or lung failure

#### Prognosis

The prognosis for TSS is better than it was in the 1980s, partly because of earlier diagnosis and treatment. In 1980 the mortality rate for women with menstrual-related toxic shock syndrome was 13 percent; as of 2005, it was 3–5 percent. The mortality rate in other patient groups, however, can run as high as 30 percent, particularly if the infection is caused by *S. pyogenes* rather than by *S. aureus*.

Most patients with toxic shock syndrome or TSLS are sent home from the hospital after a week and recover fully in two to three weeks. The condition can be fatal within a few hours, however, if it is not treated promptly. About half of patients lose their hair and nails within two or three months after the illness, but these will usually grow back on their own.

About 50 percent of people treated for TSS or TSLS will develop recurrences between four and six months after the acute illness. These recurrences are not usually as serious as the first episode but must still be treated in the hospital.

#### Prevention

The CDC recommends the following precautions to lower the risk of toxic shock syndrome or TSLS:

- Women of childbearing age should use low-absorbency tampons during their menstrual periods, change them every four to eight hours, or use sanitary napkins rather than tampons. Women who have had TSS or any serious staph or strep infection should not use tampons at all.
- People who have had nasal surgery should watch carefully for signs of infection, particularly if the nose has been packed with gauze or surgical dressings.
- Skin infections should be treated promptly.
- People who have had abdominal surgery, or women who have recently given birth, should also be monitored for signs of infection.

### WORDS TO KNOW

**Dialysis:** A process in which the blood of a patient with kidney failure is cleansed of the body's waste products by being pumped through a machine that filters the blood and then returns it to the body.

**Exotoxin:** A toxin secreted by a bacterium or other disease organism into the body tissues of an infected individual.

Sepsis: The medical term for blood poisoning.

**Shock:** A medical emergency in which the body's tissues do not receive enough blood due to problems with the circulatory system.

# The Future

TSS and TSLS are fortunately very rare illnesses. Research is currently focused on understanding why some people develop toxic shock syndrome when most people exposed to *S. pyogenes* and *S. aureus* do not. These bacteria are very common in all parts of the world and can be found in the throats or on the skin of people who are not sick. Recent studies indicate that the majority of adults have antibodies in the blood that protect them against the bacterial exotoxins that cause TSS and TSLS, but scientists do not yet understand why a small number of people lack these antibodies.

SEE ALSO Scarlet fever; Staph infection; Strep throat

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# Toxoplasmosis

#### Definition

Toxoplasmosis is an infectious disease caused by a one-celled parasitic protozoon. It gets its name from the scientific name of the parasite, *Toxoplasma gondii*. It is classified as a zoonosis because it can be transmitted from animals to people.

#### Description

Toxoplasmosis is an infectious disease caused by a one-celled parasite called *Toxoplasma gondii*. Although the parasite reproduces sexually only inside cats, it is widespread in many other animals, including rats, mice, birds, deer, and pigs. Most humans who become infected get toxoplasmosis from eating raw or undercooked meat or drinking water containing the parasite rather than from pet cats.

About 90 percent of people who have been infected by the parasite have no symptoms at all; some people develop a flu-like illness that lasts about a month; and a few people with weakened immune systems develop severe disease that can particularly affect the brain. It is also possible for a pregnant woman who is infected by the parasite for the first time during pregnancy to pass the infection to her unborn baby.

### Demographics

Toxoplasmosis is very common in the developed world, particularly in countries where people like to eat raw or undercooked meat. It is

Also Known As Toxoplasma infection

#### Cause

Protozoa (single-celled animal-like organisms)

#### Symptoms

None in most people; flulike illness in some; brain, lung, or eye damage in a few

#### Duration

About a month



Scar (center) on the retina of a patient with toxoplasmosis. © PHOTOTAKE INC. / ALAMY.

estimated that about 20 percent of the population of the United States, or 60 million people, have been infected by the parasite. Around the world, the average rate of infection is about 30 percent, but in some countries like France, the rate hovers around 75 percent because so many people prefer undercooked meat.

Toxoplasmosis is the third most common cause of fatalities from food-related illnesses in the United States; it causes about 250,000 cases each year, resulting in 5,000 hospitalizations and about 750 deaths. About 3,500 babies are born each year in the United States with toxoplasmosis.

The rate of illness is high among patients with AIDS because of their weakened immune systems. Florida has a particularly high rate of toxoplasmosis in AIDS patients; as many as 40 percent are infected. Between 1 and 5 percent of AIDS patients in the United States eventually develop encephalitis (inflammation of the brain) from toxoplasmosis.

Toxoplasmosis affects both sexes and all races equally, as far as is known.

#### **Causes and Symptoms**

The protozoon that causes toxoplasmosis has a complex life cycle but reproduces sexually only inside cats. When a cat eats a dead mouse, rat,

# **Keeping Kitty Healthy**

Although cats are carriers of the toxoplasmosis parasite, people do not get the disease from petting or playing with a pet cat, since cats do not carry the parasite on their fur. They also do not pass the parasite to humans by scratching them. People who enjoy having cats as pets do not have to give them up even if they have AIDS, are pregnant, or are being treated for cancer.

The American Veterinary Medical Association (AVMA) recommends the following steps to lower the risk of getting toxoplasmosis from a cat:

- Keep the cat indoors. In addition to lowering the cat's risk of getting the parasite by eating dead animals, keeping kitty indoors protects him or her from automobiles, coyotes and other predators, and other parasitic infections.
- Do not feed the cat raw or undercooked meat. Feed only well-cooked meat or processed cat foods.
- Scoop the cat's litter box every day. Disposing of feces as soon as possible prevents toxoplasmosis cysts from maturing.
- Pregnant women should ask another family member to clean the cat's box, or wear disposable gloves and wash the hands well in hot soapy water afterward.
- Have a sick cat checked by a veterinarian. Most cats with toxoplasmosis, like most humans, have no signs of infection at all and recover completely even without treatment.

bird, or raw meat that contains cysts (a capsule containing the parasite in its resting stage), the cysts open up within the cat's digestive tract. There they reproduce and form new cysts, which are expelled in the cat's feces. Cats shed these cysts for only one or two weeks, after which they cannot spread the infection further.

It takes the cysts in the cat's feces between one and five days to mature. This delay is the reason why cleaning a cat's litter box on a daily basis helps to prevent toxoplasmosis in humans. The parasite's cysts are hardy; they can live for a year in garden soil. They can survive freezing temperatures as low as 10°F and cannot be killed by household bleach or detergents. The cysts can, however, be killed by freezing at temperatures below 0°F ( $-18^{\circ}$ C) or by heating food containing them to temperatures above 150°F (66°C).

People can get toxoplasmosis in one of several ways: from eating uncooked meat containing cysts; by eating unwashed vegetables that have been contaminated with cysts from garden soil; by drinking water contaminated by the parasite; by failing to wash hands after handling cat feces and getting cysts into the mouth accidentally; by direct transmission from mother to child; and by receiving an infected organ by transplantation or infected blood through transfusion (very rare). People cannot get the disease from touching or being close to an infected child or adult or from petting an infected cat.

Once inside the human body, the cysts release the parasite, which forms new cysts, especially in the muscles and brain. In people with healthy immune systems, the cysts usually remain dormant for the rest of the person's life. In persons with immune systems weakened by AIDS or chemotherapy for cancer, however, the parasites may cause a flu-like illness or severe infections of the central nervous system. People with the mild form of toxoplasmosis typically have the following symptoms:

- Headache
- Fever and sore throat
- Sore aching muscles
- Swollen lymph nodes
- Night sweats

Patients with a severe toxoplasmosis infection may have:

- Lung infections resembling pneumonia
- Headache
- Loss of coordination
- Seizures
- Mental confusion and disorientation
- Blurred vision (in people with normal or weakened immune systems)

The risk of harm to an unborn baby from toxoplasmosis is highest if the mother becomes infected during the last three months of pregnancy. Toxoplasmosis can cause a pregnant woman to lose her baby before birth. The baby may also be born with seizures, disorders of the liver, or a severe eye infection. Some babies who are infected with toxoplasmosis before birth may have normal health for several years and then develop hearing loss, eye disorders, or mental retardation in the early teen years.

# Diagnosis

People are not routinely screened for toxoplasmosis because most who are infected do not develop symptoms. Those with a mild case of the disease may be thought to have an illness with similar symptoms like flu or mononucleosis. The diagnosis of toxoplasmosis can be made, however, by taking samples of the patient's blood and sending them to a laboratory for analysis.

A pregnant woman who thinks she may have been infected can be tested for toxoplasmosis by having a small sample of the fluid surrounding the baby in the womb withdrawn through a needle and tested for the parasite. This test increases the risk of losing the baby, however. The Centers for Disease Control and Prevention (CDC) advises women who are planning to become pregnant or have recently become pregnant to have a blood test for toxoplasmosis.

A patient who has symptoms of a brain disorder caused by the parasite can be evaluated by magnetic resonance imaging or a CT scan, as the infection produces characteristic patterns of damage to brain tissue that can be distinguished from brain tumors or other disorders.

#### Treatment

No treatment is needed for patients who have no symptoms of infection. Patients who have a mild case of the disease are treated with a combination of an antibiotic and a drug that was originally developed to treat malaria. People with AIDS might need to take this drug combination for an extended period to prevent symptoms from recurring. Pregnant women are given an antibiotic by itself, as the antimalarial drug can have serious side effects for both mother and child.

#### Prognosis

The prognosis of toxoplasmosis depends on the severity of the infection. Most people never know they have been infected. People with a mild form of illness usually recover completely in a few weeks. Those with severe toxoplasmosis may have lifelong eye disorders, personality changes, or seizures; about 15 percent will die from the infection.

#### Prevention

The CDC recommends the following steps to lower the risk of getting toxoplasmosis:

- Cook foods thoroughly, especially meats. Cook beef, lamb, and veal to 145°F (63°C), pork and venison (deer meat) to 160°F (71°C), and poultry to 180°F (82°C).
- Do not eat raw or undercooked meat.
- Wash knives and cutting boards in hot soapy water after use.
- Do not eat raw unwashed fruits and vegetables.
- Freeze meat for several days before cooking.
- Avoid drinking unpasteurized milk or untreated drinking water, particularly when traveling abroad.
- Wear gloves when working in the garden.
- Take proper care of pet cats (see sidebar).

# WORDS TO KNOW

**Cyst:** A capsule or sac containing a parasite in its resting stage.

Encephalitis: Inflammation of the brain.

**Protozoon (plural, protozoa):** A one-celled animal-like organism with a central nucleus

enclosed by a membrane. Many protozoa are parasites that can cause disease in humans.

**Zoonosis:** A disease that animals can transmit to humans.

#### The Future

There is no vaccine to prevent toxoplasmosis in either humans or other animals. Although an effective vaccine may be developed in the future, the best protection against toxoplasmosis at present is prevention.

SEE ALSO AIDS; Encephalitis; Food poisoning

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# Triple X Syndrome

# Definition

Triple X syndrome is a condition in which a female is born with an extra X chromosome in each cell of the body. Those who have the extra chromosome in only some of their body cells are said to have triple X mosaicism.

# Description

Triple X syndrome is caused by the presence of an extra X chromosome in the cells of a female's body. It was discovered in 1959 by Patricia Jacobs, a researcher working in a hospital in Scotland. The patient was a thirty-five-year-old woman who had undergone premature menopause.

Although some girls with triple X syndrome have learning disabilities or developmental delays, many do not. There are no unusual physical or medical disabilities associated with the syndrome; many girls with very mild symptoms may never be diagnosed. Researchers do not know why the extra copy of the X chromosome causes an increase in height and learning problems in some girls with the syndrome.

# Demographics

Triple X syndrome occurs in about one of every 900–1,000 girls born in the United States. It is estimated that between five and ten girls are born with the condition each day. As far as is known, the syndrome is equally common in all racial and ethnic groups. The only known risk factor for the syndrome is the mother's age; women who are thirty-five or older when they become pregnant are at increased risk of having a daughter with triple X syndrome.

# **Causes and Symptoms**

The most common cause of triple X syndrome occurs when an egg carrying two copies of the X chromosome is involved in conception. The reason for the extra copy in the abnormal germ cell is a genetic error called nondisjunction. Nondisjunction is a random event and is

#### Also Known As

Trisomy XXX, XXX syndrome, 47, XXX aneuploidy

#### Cause

An extra X chromosome in some or all cells of a female's body

#### Symptoms

No symptoms unique to syndrome; some girls have none at all

#### Duration

Lifelong

not caused by anything either of the baby's parents did or by any medications they may have taken.

During the normal process of germ cell formation, the paired chromosomes in the cell divide so that each daughter cell has only one member of the pair. In nondisjunction, one daughter cell gets both members of the chromosome pair and the other cell has none. If a germ cell carrying two copies of the X chromosome is fertilized by a normal germ cell from the other parent, the child will have three copies of the X chromosome. This genetic error is called a full trisomy X.

Some girls affected by triple X syndrome children have some body cells with the extra copy of the X chromosome and some body cells without the extra copy. This condition is called mosaic trisomy X. About half of women with triple X syndrome have the full trisomy X and the other half have mosaic trisomy X.

Not all girls born with triple X syndrome have noticeable symptoms of the condition, which is why they may not be diagnosed for some years. In addition, girls with the normal number of X chromosomes may also have these features that are symptomatic of triple X syndrome:

- Slightly lower weight at birth
- Taller than other girls and women in the family
- Less coordinated; somewhat higher risk of developing back problems
- Development of language skills may be slowed
- Less assertive and quieter than most girls
- Increased risk of depression
- Intelligence that may be slightly lower than that of other girls and women in the patient's family (Mental retardation is unusual.)

The mother of a girl who was diagnosed with triple X syndrome at age ten remarks that one of the major difficulties in coping with the disorder is that little has been written about the syndrome, possibly because the symptoms are so mild in many girls. Her own daughter was diagnosed as the result of learning difficulties, particularly language skills. "Although our numbers are smaller than some of the other X and Y chromosome variations, I can't help but wonder where are all the other girls? According to the scientific literature, trisomy X occurs in approximately one out of 900 to 1,000 live female births. And yet our numbers have hovered in the hundreds of individuals for a condition that affects millions worldwide....While we all hope for the best, in part because the symptoms can be so mild and there is such a small amount of literature concerning treatments, we eventually become resigned to accept that the problems we experience are just an unavoidable given that we have to deal with."

The first national conference on triple X syndrome in the United States was held in October 2006. It is hoped that regular meetings will lead to new research and better understanding of the syndrome. Girls with trisomy X go through puberty at the same age as girls with two X chromosomes and develop all the normal sexual characteristics of adult women. They are able to marry and have children, and do not need hormone treatments in order to become sexually mature. It is not yet known whether adult women with triple X syndrome undergo menopause at an earlier age than most women because the condition has not been studied long enough for researchers to follow a significant number of subjects through adult life into menopause.

### Diagnosis

Triple X syndrome may never be diagnosed if the girl has no obvious symptoms. It can, however, be detected by amniocentesis before birth. Amniocentesis is a procedure in which the doctor withdraws a small amount of fluid from the sac that surrounds the baby in the mother's uterus and examines cells from the baby's tissues for genetic abnormalities. Triple X syndrome is sometimes discovered after birth if the girl has developmental delays or learning difficulties and is given a genetic evaluation.

### Treatment

There is no specific treatment for triple X syndrome; many doctors do not consider it a major disability. Girls with the syndrome who do have learning disabilities or emotional problems are given the same types of therapy as other children with the same conditions. An individualized treatment plan can be drawn up with the help of the girl's doctor and teachers.

The most important aspect of treatment for the syndrome is love and support from the patient's family. According to a Danish researcher who started one of the first triple X support groups in the world, "If conditions at home are good, stable and stimulating...learning problems rarely have any serious consequences. However, most triple X girls need remedial teaching at school at one time or another, and it is important that they get this help if and when they need it."

# Prognosis

The prognosis for girls with triple X syndrome is generally good, particularly if their parents are loving and supportive. They are not at increased risk of other diseases as far as is known and have a normal life

# WORDS TO KNOW

**Mosaicism:** A condition in which a person has some body cells containing an abnormal number of chromosomes and other cells containing the normal number. Mosaicism results from random errors during the process of cell division that follows conception.

**Nondisjunction:** A genetic error in which one or more pairs of chromosomes fail to separate during the formation of germ cells, with the result that both chromosomes are carried to one daughter cell and none to the other. If an egg or sperm with a paired set of chromosomes is involved in the conception of a child, the child will have three chromosomes in its genetic makeup, two from one parent and one from the other.

**Trisomy:** A type of genetic disorder in which a cell contains three copies of a particular chromosome instead of the normal two.

expectancy. They are less likely to complete college than other girls, but almost all can finish high school with speech therapy or other additional help. While a few girls with triple X syndrome have some social difficulties, most are able to join the workforce, marry, and start families in adult life.

#### Prevention

Because triple X syndrome is thought to be caused by a spontaneous genetic mutation rather than an inherited genetic defect, there is no way to prevent it.

# **The Future**

It is possible that the increasing numbers of women having children in their thirties or forties will lead to an increase in the number of girls born with triple X syndrome. Little research has been done on triple X syndrome considering the numbers of girls affected by it; however, the conference hosted by the University of California, Davis, in 2006 offered some possible new directions. Papers were presented on the treatment of language difficulties, depression, anxiety, and developmental differences in girls with triple X syndrome. Another presenter discussed the use of medications as part of treatment plans for these girls.

**SEE ALSO** Down syndrome; Dyslexia; Edwards syndrome; Klinefelter syndrome; Patau syndrome

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# Trisomy 13

See Patau syndrome.

Trisomy 18 See Edwards syndrome.

Trisomy 21 See Down syndrome.



# **Tuberculosis**

# Definition

Tuberculosis, or TB, is an infectious disease caused by rod-shaped bacteria called *Mycobacterium tuberculosis*. It mostly attacks the lungs but can also infect other organs. TB has been known for centuries and was first recognized as a contagious disease in the eleventh century by Ibn Sina (980–1037), a Persian doctor. TB was not recognized as a single illness until the 1820s, however. Robert Koch (1843–1910), the German doctor considered the father of microbiology, identified *M. tuberculosis* as the cause of tuberculosis in 1882. He received the Nobel Prize in physiology or medicine for this discovery in 1905.

#### Description

Although it is no longer the leading cause of death in the United States, TB is a leading cause of infection-related death worldwide. The World Health Organization (WHO) estimates that 3 million people die of TB each year around the world.

TB is spread by droplets coughed or sneezed into the air. Other people can become infected if they breathe in these droplets. TB is not known to affect any species other than humans. After the tuberculosis bacteria enter the body, they pass down through the airway to the lungs, with one of three outcomes:

- The bacteria multiply and cause primary tuberculosis. This condition is also called active TB.
- The bacteria may become dormant (inactive) and the patient will not feel sick or be able to spread the disease. This condition is called latent TB, because the patient will test positive for the bacterium if they have a skin test. About 90 percent of people infected with TB have the latent form of the disease.
- The bacteria are dormant for a while, but then begin to multiply again and the patient feels sick. This condition is called reactivation TB. About 10 percent of people with latent TB will eventually develop reactivation TB during their lifetimes, half of these cases occur in the first two years.

A person with active TB may first notice a cough lasting three weeks or longer with bloody sputum (mucus or phlegm). The coughing or even breathing may cause pain. Night sweats and a low-grade fever usually appear. Another common symptom is loss of appetite and unintended weight loss; in fact, TB used to be called consumption because the patient's body looked as if it were being consumed, or eaten up, from within.

In addition to the lungs, tuberculosis can spread through the bloodstream and affect the spine, bones and bone marrow, joints, kidneys, muscles, and central nervous system, which might create symptoms such Also Known As TB, consumption

**Cause** Bacterium

#### Symptoms

Long-lasting cough with bloody sputum, low-grade fever, drenching night sweats, weight loss, painful breathing or coughing

**Duration** Months to years
#### Tuberculosis

Magnified image of the bacteria that cause tuberculosis. © MEDICAL-ON-LINE / ALAMY.



as severe back pain, blood in the urine, abdominal pain, swollen lymph nodes, or skin ulcers. These manifestations of TB are usually part of reactivation TB and can occur without any evidence of lung TB.

# Demographics

The frequency of active TB dropped in the United States even before the introduction of antibiotics in the 1950s, a decrease related to better living conditions and nutrition. It began to rise again in the 1980s among ethnic minorities and persons with HIV infection. In 1993 there were 25,200 cases of TB reported in the United States; that number declined again to 13,300 cases in 2007. There are about 4.4 cases of active TB in the United States for every 100,000 people. Over half these cases involve immigrants from four countries: Mexico, the Philippines, India, and Vietnam. The Centers for Disease Control and Prevention (CDC) estimates that 10–15 million people in the United States have latent TB as of 2008.

In general, it is difficult to become infected with the TB bacillus unless one lives in close long-term contact with a person with active TB. There is increased risk of infection for these groups:

- The elderly
- Those who have immune systems weakened by such diseases as AIDS, diabetes, rheumatoid arthritis, kidney disease, or Crohn disease

- Those who are malnourished or severely underweight
- Patients who have developed silicosis (a lung disease caused by breathing in rock dust produced by blasting and drilling)
- Homeless people
- People living in refugee camps or other crowded conditions
- Drug or alcohol abusers
- Patients being treated for cancer with chemotherapy, or who have received organ transplants
- Those who work or live in hospitals, nursing homes, prisons, and other institutions for long-term care
- People who come from countries with high rates of active TB

# **Causes and Symptoms**

TB is caused by a bacterium, *Mycobacterium tuberculosis*. The bacteria enter a person's respiratory tract through the nose or throat, then travel to the lungs, where they multiply within the tiny air sacs known as alveoli. In the alveoli, the bacteria are picked up by cells that carry them to nearby lymph nodes in the chest cavity. The TB bacteria can then spread into the bloodstream and other organs or tissues.

# Drug-Resistant Tuberculosis Bacteria

Beginning in the 1980s, doctors began to notice that people who failed to take medications for active TB were developing strains of the TB bacillus that are resistant to treatment. The two drugs that are considered most effective in treating TB are isoniazid (INH) and rifampin. A patient is considered to have multiple drug-resistant tuberculosis (MDR-TB) if a laboratory test shows that the bacteria in the sample are resistant to those two specific drugs. Patients diagnosed with MDR-TB must be treated for twice as long as those with ordinary TB. The drugs used to treat MDR-TB are less effective, often much more expensive, some have to be injected rather than taken by mouth, and they can produce severe side effects. Public health doctors insist that patients with MDR-TB be treated by the DOT method described in the main entry.

An even more severe form of drug-resistant TB is known as extremely drug-resistant TB or XDR-TB. It is diagnosed when a sputum test shows resistance to an injectable drug and a drug called levofloxacin as well as rifampin and isoniazid. XDR-TB is much more difficult to treat with the drugs available and has a very

The most common symptoms of active TB (that is still limited to the lungs) include:

- A cough that lasts longer than three weeks
- Pain when coughing or even breathing
- Loss of appetite and weight loss
- Pale complexion
- Fatigue

- Wheezing
- · Low-grade fever and night sweats
- Clubbing of the fingers or toes

### Diagnosis

The diagnosis of TB can be difficult because the disease does not produce symptoms immediately even with an active infection. People exposed to a person with active TB and are at high risk to become infected should check with their doctor even if they do not feel sick. The doctor will perform a physical examination. Some people with active TB develop noises in the lungs known as crackles that can be heard through a stethoscope. The doctor will also feel the lymph nodes in the patient's neck and look for such other signs as clubbing of the finger tips.

A skin test known as the Mantoux test (also called the PPD test) is used to screen patients for infection with TB. The doctor injects a fluid derived from the TB bacteria under the top layer of the patient's skin. If the person has been infected with TB, a raised hard flat area will develop within forty-eight to seventy-two hours at the site of the injection. The patient's risk factors are used to determine how large the bump must be to be considered a positive reaction. A newer test is a blood test approved by the Food and Drug Administration (FDA). Called the QuantiFERON-TB Gold (QFT) test, it detects the presence of TB bacteria in the patient's blood and gives results in a day. As of 2008, it was not yet available in all parts of the United States, however.

A patient who tests positive on either the Mantoux or the QFT test will then be given a chest x ray and a sputum test. A sample of sputum is sent to a laboratory where it is cultured, to see whether the person has active TB and to see whether the TB bacteria respond to standard antibiotics. If not, the patient has multidrug-resistant TB (MDR-TB) and will need special treatment.

# Treatment

Patients who are found to have latent TB may be treated with a drug called isoniazid or INH. This drug is given to destroy dormant bacteria so that they cannot reactivate in the future. The patient usually takes INH for six to nine months.

Patients with active TB are treated with INH plus a combination of three other drugs to make sure that all the TB bacteria are destroyed. The doctor may change one of the drugs during therapy if it turns out that the patient has a form of the TB bacillus resistant to that particular drug. Drug treatment for TB is a long-term process that can take anywhere from six months to a year for non-drug resistant TB; MDR-TB may require two full years of drug therapy. The patient may be admitted to the hospital for the first two weeks of treatment or until tests show that they are no longer contagious.

It is very important for patients with active TB to take all their drugs exactly as directed for the full course of therapy even though they will usually start to feel better in a few weeks. If they do not, TB bacteria may survive and develop resistance to the drugs. MDR-TB is much more difficult to treat and is very dangerous. To make sure that patients take their medications correctly, some doctors or clinics use an approach called directly observed therapy or DOT. In DOT, a nurse or other health care worker gives the patient their drugs in the clinic so that they do not have to remember to take their medications at home.

Some patients with severe MDR-TB may need surgery as well as medications. Removing part of an infected lung reduces the number of active bacteria in the patient's body; it may also increase the effectiveness of drug therapy.

### Prognosis

The prognosis of active tuberculosis depends on the patient's age, overall health, and whether he or she has a drug-resistant strain of TB. About 4 percent of patients in the United States with active TB die from the disease; this is much lower than the 50 percent death rate in the 1920s. In general, people with MDR-TB have a worse prognoses than those who do not; patients with HIV infection as well as MDR-TB have poor prognoses.

#### Prevention

Tuberculosis is a preventable disease. In addition to diagnosing and treating people with latent TB before they develop active infection, people can lower their risk of getting TB in several ways:

- Keep the immune system healthy.
- Get an annual skin test if one is frequently exposed to TB or if one has a weakened immune system.

# WORDS TO KNOW

**Alveoli (singular, alveolus):** Tiny air sacs in the lungs where the exchange of oxygen for carbon dioxide in the blood takes place.

- **Clubbing:** Thickening of the tips of the fingers or toes.
- **Directly observed therapy (DOT):** Treatment in which nurses or health care workers administer

medications to patients in a clinic or doctor's office to make sure that the patients take the drugs correctly.

Latent: Referring to a disease that is inactive.

**Sputum:** Mucus that comes up when a person coughs.

- Get treatment for latent TB if infected.
- Patients diagnosed with active TB should stay home, avoid close contact with others, and cover their mouths when they sneeze or cough.
- A vaccine against TB, known as BCG, is widely used in Europe. It is not very effective in adults, however, and is not widely used in the United States because it can cause a false-positive result on the Mantoux test.

# The Future

Before the 1980s, it was hoped that tuberculosis could be wiped out completely. The emergence of MDR-TB in the 1980s, however, destroyed that hope. In 1993 WHO declared the resurgence of TB as a global health emergency. Researchers are trying to develop a more effective vaccine than BCG and find new antibiotics to treat MDR-TB and XDR-TB. They are also testing fixed-dose combination tablets of anti-TB drugs to simplify patients' medications.

SEE ALSO AIDS; Pneumonia

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# **Turner Syndrome**

# Definition

Turner syndrome is a genetic disorder in which the cells in a girl's body are missing all or part of one of the two X chromosomes. The syndrome occurs only in girls. It is named for Henry Turner (1892–1970), a doctor in Oklahoma who first described the syndrome at a conference in 1938.

### Description

Turner syndrome is a genetic disorder in which a girl is missing one or part of one of the two X chromosomes that define a person's sex as female. Women with Turner syndrome are shorter than average and cannot have children because their ovaries do not develop normally. They may also have some abnormal physical features, such as extra skin around the neck that gives the neck a webbed appearance; swollen hands Also Known As Ullrich-Turner syndrome, monosomy X, TS

#### Cause

Absent or damaged X chromosome

#### Symptoms

Growth problems; infertility; kidney or heart disorders; learning difficulties

Duration Lifelong

#### **Turner Syndrome**

Image of the set of chromosomes of a patient with Turner syndrome. Women with the disease have only one X chromosome. GJLP / PHOTO RESEARCHERS, INC.



and feet; heart or kidney problems; and scoliosis (abnormal sideways curvature of the spine). They are also at increased risk of high blood pressure and osteoporosis (brittle bones) in later life.

Turner syndrome may result either from a complete lack of the second X chromosome—a condition called monosomy X—or from damage to or partial rearrangement of the second X chromosome. About half of all women with Turner syndrome have monosomy X; two-thirds of these patients lack the father's X chromosome and the remaining third lack the mother's X. The other half of patients with Turner syndrome have a damaged or incomplete second X chromosome.

Although geneticists are not certain how many genes on the X chromosome are responsible for some of the characteristics of the syndrome, they have identified one gene known as SHOX, which is responsible for the production of a protein involved in bone growth. A missing or abnormal SHOX gene is thought to cause the short stature and skeletal abnormalities of women with Turner syndrome.

# **Demographics**

Turner syndrome occurs in one in every 2,000–2,500 newborn girls in the United States. Most female fetuses with Turner syndrome, however, do not survive until birth; doctors estimate that 98 percent die before birth or are miscarried. The rate of Turner syndrome appears to be the same in all countries around the world and in all races and ethnic groups. No risk factors have been identified that increase the likelihood of parents having a daughter with the syndrome; as far as is known, the genetic errors that cause the disorder occur at random.

# **Causes and Symptoms**

Turner syndrome involves genetic errors that occur during the formation of germ cells (eggs and sperm) or during cell division shortly after the egg is fertilized by the sperm. The more common form of the syndrome occurs when an egg or sperm lacking an X chromosome is involved in conception. The reason for the missing copy in the abnormal germ cell is a genetic error called nondisjunction. During the normal process of germ cell formation, the paired chromosomes in the cell divide

so that each daughter cell has one member of the pair. In nondisjunction, one daughter cell gets both members of the chromosome pair and the other cell has none. If a germ cell lacking an X chromosome is fertilized by a normal germ cell from the other parent, the child will have only one X chromosome. This genetic error is called monosomy X.

Some cases of Turner syndrome occur in girls who have some body cells with two X chromosomes and some body cells without the second X. This condition is called mosaic Turner's. It is thought to result from random errors in cell division during the early stages of fetal development.

The symptoms of Turner syndrome include:

- Short height in adult life. The average height of an adult woman with Turner syndrome is 4 feet 7 inches (1.4 meters).
- Flat, shield-shaped chest with widely spaced nipples.
- Wide neck with webbing (extra skin) at the base.
- Swelling of the hands and feet.

# An Academy Award Winner

Linda Hunt (1945–), is a film and television actress who won an Oscar in 1983 for her portrayal of a male Chinese-Australian photographer in a film called *The Year of Living Dangerously*. She is the first star to win an Academy Award for playing a character of the opposite sex.

Diagnosed with Turner syndrome in adolescence, Hunt is short for an adult woman at 4 feet 9 inches (1.45 meters). She has a rich and deep speaking voice that has made her a popular choice as a narrator for such television series as *American Experience* and *California and the American Dream*. One of Hunt's bestknown film roles is the voice of Grandmother Willow in the 1997 Disney animated feature *Pocahontas*.

- Kidney abnormalities, including a single horseshoe-shaped kidney on one side of the body or poor blood flow to the kidneys.
- Low hairline and low-set ears.
- Drooping eyelids.
- Turned-out elbows.
- Frequent ear infections and hearing loss.
- Abnormalities in the structure of the heart. These may include damaged heart valves or a weakened aorta, which is the large artery that leaves the left side of the heart and carries oxygenated blood to all parts of the body. Women with Turner syndrome are at increased risk of a ruptured aorta.
- Hypothyroidism (low levels of thyroid hormone). About a third of women with Turner syndrome develop Hashimoto's disease.
- Increased risk of both type 1 and type 2 diabetes.
- Absence of menstrual periods and inability to have children.
- Specific learning difficulties, most commonly with mathematics and with spatial perception.

# Diagnosis

Turner syndrome can be detected by amniocentesis before birth. Amniocentesis is a procedure in which the doctor withdraws a small amount of fluid from the sac that surrounds the baby in the mother's uterus and examines cells from the baby's tissues for genetic abnormalities. It may also be diagnosed after the baby is born when the doctor and parents notice such features as a webbed neck, broad flat chest, swollen hands and feet, and heart defects. The diagnosis can be confirmed by a karyotype, which is a blood test used to obtain cells for examination of their chromosomes under a microscope.

In some cases Turner syndrome is not diagnosed until the girl's growth slows down in early childhood or until she fails to reach puberty. At that time that doctor may order a karyotype (a photomicrograph of the chromosomes in a single human cell).

### Treatment

Turner syndrome cannot be cured because it is a genetic condition. However, growth hormone started early and estrogen replacement treatment in adolescence can help girls with the disorder grow to a normal height and achieve normal sexual development. Early diagnosis of Turner syndrome is important because growth hormone treatment should be started in childhood. Estrogen replacement therapy is not started until the girl is twelve to fifteen years old, as it will slow down her growth in height if given earlier. Girls with Turner syndrome should continue to receive estrogen until they reach the normal age of menopause in order to prevent osteoporosis.

Patients with Turner syndrome require careful monitoring throughout life for such possible complications as recurrent ear infections and possible hearing loss; kidney or thyroid disorders; heart problems; diabetes; and high blood pressure. They should have annual blood tests as part of routine physical checkups. In addition, they should watch their weight carefully because obesity increases their already high risk of diabetes.

Although women with Turner syndrome are infertile because their ovaries do not develop normally, they can become pregnant with the help of assisted reproductive techniques. They should, however, consult a heart specialist before considering pregnancy because of the risk of aortic rupture or other heart complications.

Most girls with Turner syndrome have normal intelligence, and many are intellectually gifted. Those who have difficulty with mathematics or related areas of study may need special tutoring in those fields. In general, however, TS should not interfere with patients' ability to complete schooling and be successful in a wide variety of occupations. Several studies have shown that many women with Turner syndrome have higher-than-average educational achievements.

#### Prognosis

The prognosis of Turner syndrome depends partly on whether the patient has monosomy X, a damaged second X chromosome, or mosaic Turner syndrome. In general, patients with monosomy X have a greater chance of developing heart or kidney problems than those with other forms of Turner syndrome and require more careful monitoring of their health. Most patients have an overall good prognosis with a slightly shorter than average life expectancy.

#### Prevention

Since Turner syndrome is thought to be caused by a random genetic mutation rather than an inherited genetic defect, there is no way to

# WORDS TO KNOW

- **Karyotype:** A photomicrograph of the chromosomes in a single human cell. Making a karyotype is one way to test for genetic disorders.
- **Monosomy:** A type of genetic disorder in which a cell contains only one copy of a particular chromosome instead of the normal two.
- **Mosaicism:** A condition in which a person has some body cells containing an abnormal

number of chromosomes and other cells containing the normal number. Mosaicism results from random errors during the process of cell division that follows conception.

**Nondisjunction:** A genetic error in which one or more pairs of chromosomes fail to separate during the formation of germ cells, with the result that both chromosomes are carried to one daughter cell and none to the other.

prevent it. Parents of a daughter with Turner syndrome are unlikely to have other daughters with the disorder.

#### The Future

Turner syndrome is not likely to become more common in the general population in the future. In terms of treatments, as of 2008 the National Institutes of Health (NIH) was recruiting patients with Turner syndrome for clinical trials to evaluate the long-term effects of growth hormone therapy and estrogen replacement therapy. Research is ongoing to look for other genes on the X chromosome that may be involved in Turner syndrome.

**SEE ALSO** Hashimoto disease; Osteoporosis; Scoliosis; Triple X syndrome

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# U



Genetic Infection

Injury

Multiple

Other

Unknown

Also Known As UC, inflammatory bowel disease, IBD

#### Cause

Unknown; possibly genetic factors combined with environmental triggers

#### Symptoms

Cramping in lower abdomen, diarrhea, bloody diarrhea, bleeding from rectum, fever, weight loss

Duration

Years

# U

# Ulcerative colitis

# Definition

Ulcerative colitis, or UC, is a form of inflammatory bowel disease or IBD. Unlike Crohn disease, which is another form of IBD, ulcerative colitis affects only the large bowel or colon. It does not affect the small intestine or other parts of the digestive tract. Ulcerative colitis that is limited to the rectum (the lowermost portion of the colon) is sometimes called proctitis. Ulcerative colitis that affects the entire colon, from the rectum to the point where the colon joins the small intestine, is called pancolitis.

It is important to distinguish IBD from irritable bowel syndrome, or IBS. IBS does not involve inflammation of the bowel and is a less serious disease than IBD.

# Description

Ulcerative colitis is a disorder of the digestive tract that usually begins slowly rather than suddenly. In response to inflammation of the tissues lining the colon, small sores or ulcers form on the surface of these tissues and produce pus and bloody mucus that the body then expels. The patient typically experiences cramps in the lower abdomen and bloodor mucus-streaked diarrhea several times a day. In the early stages of the disease the patient notices that the stools gradually become looser. As the



View of the colon of an elderly woman with ulcerative colitis. Yellow areas show inflammation. DAVID M. MARTIN, M.D. / PHOTO RESEARCHERS, INC.

disease progresses, the patient may have more frequent stools and a more urgent need to defecate.

Ulcerative colitis does not affect all patients with equal severity. About half of patients with UC have only mild diarrhea (fewer than four stools per day) and cramping whereas the other half may have six or more stools per day with fever, anemia, and complications that affect the eyes or other organs. In addition, the symptoms of ulcerative colitis often come and go; periods of few or no symptoms are called remissions.

A potentially life-threatening but rare form of ulcerative colitis is known as fulminant colitis. In fulminant colitis, the patient has ten or more bowel movements per day, involvement of the

entire colon, and the potential for rupture of the colon.

Ulcerative colitis differs from Crohn disease in two important respects: first, the affected portion of the bowel is continuous; there is no pattern of alternation between diseased portions of the bowel and normal portions. Second, unlike Crohn disease, ulcerative colitis is usually limited to the innermost layer of tissue lining the colon, whereas Crohn disease can penetrate to deeper layers of intestinal tissue.

# **Demographics**

Ulcerative colitis affects between 500,000 and 700,000 people in the United States, with another 500,000 suffering from Crohn disease. Ulcerative colitis is almost entirely a disorder of older adolescents or adults, with two peak age groups: people between fifteen and thirty years of age, and people between fifty and seventy. It can, however, occur in other age groups.

In the developing world, UC primarily affects the upper classes. It is less common in Asia and Africa than in Europe and North America. In the United States, it is more common among Caucasian Jews of Eastern European ancestry than among other racial and ethnic groups, and more common in the northern states than in other parts of the country. Ulcerative colitis is slightly more common among women than men; there are three females for every two males affected. Having a family member with ulcerative colitis increases a person's risk of developing the disease, but only slightly. An identical twin of a patient diagnosed with UC has a 10 percent chance of developing it.

# **Causes and Symptoms**

The causes of ulcerative colitis are not completely understood. The most widely accepted theory holds that the disease is caused by the patient's immune system mistaking bacteria that normally live in the intestine as foreign substances that must be attacked. When the immune system overreacts, white blood cells move in large numbers to the intestines, where they accumulate and eventually cause swelling and destruction of tissue.

Another theory holds that ulcerative colitis is an inherited bowel disorder. There are certain regions of the human genome on chromosomes 1, 3, 5, 6, 12, 14, 16, and 19 that have been linked to UC; however, no single gene from any of these regions has been shown to be consistently associated with ulcerative colitis. Researchers think that a combination of several genes is necessary to trigger the disorder.

At one time it was thought that diet or emotional stress played a role in the development of UC, but these theories are no longer accepted by most doctors. In contrast to Crohn disease, smoking does not increase a person's likelihood of developing ulcerative colitis.

The most common symptoms of ulcerative colitis are bloody or mucus-streaked diarrhea, abdominal cramps, rectal bleeding, and fever or unintended weight loss. Some patients also experience a feeling of fullness in the rectum even when it is empty; night sweats; nausea or vomiting; fatigue; and loss of appetite.

It is not known why some patients with UC have symptoms in other organs or parts of the body, although researchers think that these complications are the result of inflammation triggered by the patient's immune system. Complications outside the digestive tract may include:

- Inflammation of the iris of the eye
- · Pains in the joints
- Mouth ulcers
- Liver disease
- Osteoporosis
- Skin rashes

# Diagnosis

The diagnosis of ulcerative colitis can be complicated because of the need to rule out Crohn disease, irritable bowel syndrome, and intestinal infections. Most primary care doctors will refer the patient to a gastroenterologist, who is a doctor who specializes in diagnosing and treating disorders of the stomach and intestines.

The gastroenterologist will take a history of the patient's symptoms and order one or more of the following tests:

- Blood tests. These may be ordered to check for anemia or signs of infection. A high white blood cell count typically indicates infection somewhere in the patient's body.
- Stool sample. This test can be done to rule out intestinal parasites or bacteria or viruses that may be causing bloody stools.
- Sigmoidoscopy or colonoscopy. A sigmoidoscope is a flexible lighted tube that can be inserted into the rectum and used to examine the last two feet of the colon. A colonoscope is a long flexible tube attached to a video camera and monitor that allows the doctor to examine the entire length of the patient's colon and rectum. The patient must take a laxative the night before a colonoscopy to cleanse the bowel and may be given a sedative in the doctor's office to make them more comfortable. The doctor will remove a small sample of tissue for biopsy; this is done to rule out Crohn disease.
- Barium enema. A barium enema is an enema that contains a chalky substance that coats the lining of the rectum and upper colon. X-ray photographs are then taken as the patient lies on a table.
- Computed tomography (CT) scan. The CT scan is an imaging tool to view internal organs.

# Treatment

There is no cure for ulcerative colitis. Treatment of the disease depends on the severity of the patient's symptoms and the amount of the bowel that is involved. Some patients do well with one or more drugs given to reduce inflammation and encourage remission. There are three groups of drugs commonly prescribed for ulcerative colitis:

• Aminosalicylates. These include drugs like sulfasalazine and mesalamine. They can be taken by mouth or as rectal suppositories, depending on the location of the inflammation.

- Corticosteroids. These include prednisone and hydrocortisone. Steroid medications are effective in reducing inflammation but are usually given only for short periods of time because they have potentially serious side effects.
- Azathioprine and mercaptopurine. These are drugs that work to lower inflammation by suppressing the patient's immune system.

Patients who are having a severe attack of ulcerative colitis may need to be hospitalized for treatment of dehydration and blood loss. They may need a special diet for a few weeks or intensive treatment with steroid medications to bring down the inflammation.

Patients at risk of fulminant colitis or who do not respond to medications can be treated with surgery to remove the inflamed portion of the colon. The surgeon can remove the entire colon and rectum and create an artificial opening to the outside of the abdomen called a stoma. The patient's digestive wastes are passed into an ostomy bag attached to the stoma with adhesive. A newer surgical technique involves the creation of a pouch inside the abdomen by joining the lower end of the small intestine to the inside of the rectum and the anus. The patient may still have watery or loose bowel movements but will not need to wear an ostomy bag.

#### Prognosis

In general, patients with proctitis have a milder course of the disease than those whose colitis affects a larger portion of the colon; they are also more likely to have long periods of remission. Between 25 and 40 percent of patients eventually need surgery for the disease. Ulcerative colitis does not, however, appear to shorten a person's life span.

Patients with ulcerative colitis have an increased risk of colorectal cancer except for those with proctitis only. About 5 percent of patients will eventually develop colon cancer. The risk increases after 10 years of the disease, particularly if the entire colon is involved. Most doctors recommend having a colonoscopy eight years after diagnosis of UC and every one to two years after that to check for signs of colorectal cancer.

#### Prevention

There is no known way to prevent ulcerative colitis, because the causes of the disorder are not yet known.

# WORDS TO KNOW

<b>Fulminant:</b> Referring to any disease or condition that strikes rapidly and is severe to the point of being life-threatening.	<b>Rectum:</b> The lowermost portion of the large intestine, about 6 inches (15.2 centimeters) long in adults.
<b>Gastroenterologist:</b> A doctor who specializes in diagnosing and treating diseases of the digestive system.	<b>Remission:</b> A period in the course of a disease when symptoms disappear for a time.
<b>Pancolitis:</b> Ulcerative colitis that affects the entire colon.	<b>Stoma:</b> An opening made in the abdomen fol-
<b>Proctitis:</b> The medical term for ulcerative colitis limited to the rectum.	lowing surgery for digestive disorders that allows wastes to pass from the body.

### The Future

Ulcerative colitis is not likely to become more common in the general American population. Current research is focused on learning more about the role of the immune system in triggering the disease and the effectiveness of newer drugs in treating symptoms.

SEE ALSO Colorectal cancer; Crohn disease; Irritable bowel syndrome

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# **m** Ulcers

# Definition

An ulcer is an open sore that forms in the skin or the lining of the mouth or digestive tract (esophagus, stomach, and intestines). It may be caused by pressure that cuts off circulation in the skin over bony parts of the body in bedridden patients; by complications of diabetes; or by infections that inflame the skin or mucous membranes (soft tissues that line the digestive and respiratory tracts) and lead to erosion of tissue. Ulcers can occur in different parts of the body. Ulcers in the digestive tract are called peptic ulcers.

# Description

Peptic ulcers are sores in the soft tissues that line the esophagus, the stomach, and the duodenum (the first part of the small intestine). They develop when tissues have been eaten away by stomach acid and digestive juices. As the tissues break down, the person experiences a burning or gnawing pain in the upper part of the abdomen. Although most peptic ulcers are small in size, the high acidity of the digestive juices in the stomach can cause intense pain when the acid in the juices touches the open sores.

# Demographics

Peptic ulcer disease, or PUD, is a very common disorder of the digestive system in developed countries. In the United States, treatments for PUD

#### Also Known As

Bedsores, canker sores, diabetic ulcers, peptic ulcer disease, PUD

#### Cause

Infections or loss of blood circulation in affected parts of the body

#### Symptoms

Open painful areas in the mouth, digestive tract, skin, or eyes; heartburn, nausea, chest pain, vomiting blood, dark tarry stools (peptic ulcers)

#### Duration

Days to months



A human stomach with a gastric ulcer that perforated, or burned a hole through the wall of the stomach. CNRI / PHOTO RESEARCHERS, INC.

account for 10 percent of the health care costs for digestive diseases. About 4.5 million Americans have PUD at any given time, and one person in ten will develop a peptic ulcer at some point in his or her life. There are approximately 350,000–500,000 new cases of PUD and more than 1 million ulcer-related hospitalizations each year in the United States. About 6,000 people die each year of complications from peptic ulcers.

Peptic ulcers are largely a disease of adults; they are relatively rare in children and adolescents. The location of peptic ulcers varies somewhat according to age and sex, however; duodenal ulcers are more common in adults between the ages of thirty and sixty and are twice as common in men as in women. Gastric (stomach) ulcers are more common in adults over sixty and are more common in women than in men.

Peptic ulcers are somewhat more common among African Americans and Hispanics than among Caucasians and Asian Americans in the United States.

# **Causes and Symptoms**

Most peptic ulcers are caused either by infection by a bacterium called *Helicobacter pylori* or by heavy use of over-the-counter nonsteroidal antiinflammatory drugs, or NSAIDs. About 61 percent of all duodenal ulcers and 63 percent of gastric ulcers are caused by *H. pylori*. This spiral-shaped bacterium is found in the digestive tracts of about 50 percent of the world's population; however, most people who have *H. pylori* in their digestive systems do not have any ulcer symptoms.

In about 20 percent of people infected with *H. pylori*, the bacterium is able to weaken the mucous tissues that protect the deeper layers of stomach and intestinal muscle. Once *H. pylori* burrows through the mucous tissues, the acid in the digestive juices irritates the tissues further and eventually produces an ulcer.

The next most common cause of peptic ulcers is NSAIDs. These are pain relievers that include such drugs as aspirin, ibuprofen, naproxen, and newer anti-arthritis drugs like celecoxib. NSAIDs can weaken the protective lining of the stomach, causing peptic ulcers directly or making it easier for *H. pylori* to penetrate the lining. Elderly people are at increased risk of PUD caused by NSAIDs because many of them take these drugs routinely for arthritis or headaches.

Before *H. pylori* was identified as a major cause of PUD in 1982, doctors thought that ulcers were caused by eating spicy foods or having a stressful job, but these theories are no longer accepted.

A small proportion of peptic ulcers are caused by:

- · Stress related to recovery from severe burns
- Zollinger-Ellison syndrome, a rare disease that produces tumors that cause the stomach to secrete abnormally high levels of acid
- Excessive alcohol drinking
- Smoking
- · Being treated with radiation therapy for cancer

The most noticeable symptom of PUD is a burning or gnawing pain behind the breastbone or in the upper middle portion of the abdomen several hours after a meal. The pain is often worse in the early morning or at night, and can last anywhere from a few minutes to several hours. It may also come and go over a period of a few days or weeks. The pain of a peptic ulcer is usually relieved by eating a small amount of bland food, by taking antacids, or by vomiting. Other symptoms of PUD include:

- Nausea and vomiting
- Black or tarry-looking stools
- · Vomiting up blood
- Chest pain
- Unintended weight loss
- Loss of energy

# Diagnosis

Peptic ulcers are usually diagnosed by a series of tests:

- Upper GI (gastrointestinal) series: This is the medical term for an imaging study in which the patient drinks a chalky liquid containing barium, a substance that coats the lining of the esophagus, stomach, and small intestine and shows up on an x ray. Most ulcers, though not all, will be visible on the x-ray photograph.
- Endoscopy. An endoscope is a thin, flexible tube with a light and a miniature camera at one end. The doctor threads the endoscope down through the esophagus into the stomach. The instrument can be used to remove a small sample of the stomach lining for biopsy as well as to take photographs of the inside of the digestive tract.
- Tests for *H. pylori*. If the doctor detects a peptic ulcer through an upper GI series or an endoscopy, further testing must be done to see whether the ulcer is caused by *H. pylori*, because the treatment is different from that for ulcers caused by NSAIDs or other factors. *H. pylori* can be detected by a blood test that measures antibodies to the bacterium, by a biopsy of stomach tissue removed during an endoscopy, or by a breath test. The breath test is done by having the patient drink a solution of urea (a watery compound containing nitrogen) combined with radioactive carbon. If *H. pylori* is present in the patient's stomach, the bacterium will break down the urea and release the carbon, which the patient will exhale in the form of carbon dioxide. The patient breathes into a plastic bag about thirty minutes after drinking the urea and carbon solution, and the amount of carbon dioxide in the breath is measured.

# Treatment

Treatment of PUD depends on whether the ulcer is caused by *H. pylori*, by NSAIDs, or by lifestyle factors. Ulcers caused by NSAIDs, by smoking, or by heavy drinking can be treated by quitting smoking, cutting back on alcohol, and by substituting other painkilling medications for NSAIDs. These ulcers can also be treated by prescription medications that coat the lining of the stomach until the ulcer heals, or other medications that block the production of stomach acid.

Peptic ulcers caused by *H. pylori* are treated by a combination of two or more medications. The goal of this type of therapy is to eliminate the bacterium.

- Antibiotics. Two or three different antibiotics are given together to eliminate *H. pylori* because one antibiotic alone will not kill the bacterium. There are now two combination drugs available for *H. pylori* infection, each drug combining two antibiotics plus a drug to block acid production. These combination medications are called Prevpac and Helidac.
- Pepto-Bismol plus antibiotics. The combination of Pepto-Bismol with two antibiotics works for 80–95 percent of people infected with *H. pylori*.
- Medications that block the production of stomach acid.
- Antacids. Antacids work by neutralizing stomach acid rather than preventing its production; they also provide rapid relief from the pain of a peptic ulcer.

Most peptic ulcers respond to treatment with medications. In rare cases, the doctor may recommend surgery to treat the ulcer. The most common surgical procedures done to treat PUD are cutting part of the nerve from the brain to the stomach that controls the production of stomach acid and removing the portion of the stomach that produces a hormone that stimulates acid production.

# Prognosis

The prognosis for peptic ulcers is excellent; almost all patients can be treated successfully by avoiding NSAIDs, taking appropriate medications, and getting treatment for *H. pylori* infection.

# Prevention

People can do several things to lower their risk of peptic ulcers:

- To avoid infection with *H. pylori*, people should stay away from contaminated food and water and observe strict standards of personal hygiene. They should wash hands carefully with warm water and soap every time they use bathroom, change a diaper, and before and after they prepare food.
- They should use lower doses of NSAIDs, use acetaminophen (Tylenol) in place of NSAIDs, or consult their doctor about pain relievers that are easier on the stomach.

# WORDS TO KNOW

**Duodenum:** The first part of the small intestine. **Gastric:** Related to the stomach. **Peptic ulcer:** The medical term for an ulcer in the digestive tract.

- They should quit smoking.
- They should lower the amount of alcohol, tea, and coffee that they drink.

# The Future

Peptic ulcers caused by *H. pylori* appear to be decreasing in frequency in the United States, while those caused by NSAIDs are increasing, particularly among the elderly. This increase is not surprising given the aging of the general population and the number of age-related health problems that are treated with NSAIDs. Considerable research is presently directed toward finding pain-relieving drugs that would be easier on the digestive system. One experimental drug in clinical trials is a combination of ibuprofen, an NSAID, with a drug that blocks acid production. The researchers are investigating whether the combination medication will reduce the rate of ulcer development in patients who need a daily dose of ibuprofen over long periods of time.

**SEE ALSO** Alcoholism; Burns and scalds; Canker sores; Diabetes; Gangrene; Irritable bowel syndrome; Smoking; Stress

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# Urinary tract infection

# Definition

Urinary tract infections, or UTIs, are disorders caused by infections, mostly bacterial but sometimes fungal or viral, of the urinary bladder, the urethra, the ureters, or the kidneys themselves. Infection of the bladder by itself is called cystitis; infection that has moved upward to the kidneys from the bladder or ureters is called pyelonephritis. UTIs can be caused by several different types of bacteria, but the most common offenders are bacteria that live in the intestines and get into the urinary tract because of poor cleansing after using the toilet. Other organisms that may be involved in a UTI include those that cause herpes, gonorrhea, and other sexually transmitted diseases.

# Description

The human urinary tract is usually free of bacteria, as is the urine it produces. Bacterial infections usually start when bacteria from the outside of

#### Also Known As

UTI, bladder infection, cystitis

#### Cause

Bacteria in the kidneys, bladder, ureters, or urethra

#### Symptoms

Fever; pain on urination; need to urinate frequently; cloudy, smelly, or reddish urine

#### Duration

Three to ten days in most cases

#### Urinary tract infection

Computer image of bacteria (blue) in the urinary tract. Normally no bacteria are in the urinary tract. DAVID MACK / PHOTO RESEARCHERS, INC



the body enter through the urethra, a thin tube that connects the urinary bladder in the lower abdomen to the outside. The urethra in adult women is only about 1.5–2 inches (3.8–5.1 centimeters) long, whereas the adult male urethra is about 8 inches (20.3 centimeters) long. This difference helps to explain why adult women are the group at greatest risk of UTIs. Bacteria from the area around the anus or the vagina can enter the female urethra during sexual intercourse, or they can be introduced into the opening of the urethra by wiping from back to front after using the toilet. In men, bacteria are most likely to get into the urethra following surgery or insertion of a urinary catheter.

Once the bacteria have entered the body through the urethra, they travel upward into the bladder, a hollow muscular organ in the lower abdomen that collects and stores urine. In the bladder, the bacteria may cause an inflammation known as cystitis, which is characterized by pain on urination, a need to urinate frequently, and cloudy, bloody, or foulsmelling urine.

If the infection is not treated, it may spread upward past the bladder through the ureters into the kidneys. The ureters are two tubes about 10 inches (25.4 centimeters) long in human adults that connect the kidneys to the bladder. A bacterial infection of the kidneys is more serious than a bladder infection because of the possibility of long-term damage to the kidneys. Patients with pyelonephritis may experience vomiting, severe pain in the abdomen, night sweats, and a high fever as well as painful urination.

# Demographics

UTIs are largely an adult disorder, although between 1 and 2 percent of children are affected by them. According to the American Urological Association, urinary tract infections are responsible for more than 7 million visits to doctors each year in the United States and about 5 percent of all visits to primary care doctors. About 40 percent of women and 12 percent of men will experience at least one UTI during their lifetime. Although UTIs are less common in boys and men than in girls and women, they are also more serious when they do occur.

As far as is known, race or ethnicity does not affect a person's risk of UTIs. Factors that do increase risk, however, include:

- Female sex. The female urethra is only a quarter as long as the male urethra, which makes it easier for bacteria to enter the female bladder.
- Sexual activity. Sexual intercourse can irritate the opening of the urethra in women and allow bacteria to enter.
- Pregnancy.
- Age. Older men are more likely to develop UTIs because the swelling of the prostate gland may make it more difficult to empty the bladder completely. Women past menopause have an increased risk of UTIs because of the weakening of the muscles surrounding the bladder.

# UTIs in Children

Most people think of urinary tract infections (UTIs) as an adult problem, but between 1 and 2 percent of children develop UTIs. These can be serious, because small children are much more likely than older children or adults to suffer lasting kidney damage from UTIs, and children may be too young to tell an adult when and where they are having pain; they may have only a fever, smelly urine in a diaper, diarrhea, or no symptoms at all. It is best to take a small child to the doctor if he or she is running a high fever and there is no other obvious explanation for it such as an ear infection or a cold.

As with an adult, the diagnosis of a UTI in a small child is based on a urine sample. After the infection clears, the doctor may recommend additional tests to check for abnormalities in the structure of the child's ureters or urethra. In some cases children get recurrent UTIs because they are born with abnormally narrow passages in one or more parts of their urinary tract, which can prevent the normal passage of urine out of the body and encourage the growth of bacteria in the urine. Doctors usually recommend x-rays or other imaging studies for children with UTIs when the child is:

- A girl over age five who has had two or more UTIs
- A boy of any age who has his first UTI
- Younger than five
- · Running a high fever
- Women who use diaphragms as a method of birth control. The rigid frame on the outside of a diaphragm can irritate the urethra or put pressure on it.
- People with diabetes.

- People who have become dehydrated or are not drinking enough fluids.
- People with anatomical abnormalities of the urinary tract.
- People with a history of kidney stones.
- People who have had surgery that requires the placement of a urinary catheter.

# **Causes and Symptoms**

The cause of urinary tract infections is the introduction of bacteria into the urinary tract. If the bacteria are not washed out by urine, they may multiply in the tissues lining the urethra and the bladder, causing inflammation, fever, pus in the urine, and other symptoms of infection. It is possible for a person to have a UTI without noticeable symptoms of illness; however, most people will feel sick fairly rapidly after the infection begins.

The symptoms of a bladder infection include:

- Burning or painful urination
- A low fever (below 101°F/38.3°C)
- Feeling a need to urinate frequently
- · Cloudy, bloody, or foul-smelling urine
- Need to urinate at night
- A feeling of pressure in the lower part of the pelvis

If the infection has spread to the kidneys, the patient may also have:

- A high fever (over 101°F/38.3°C) and chills
- Night sweats
- Severe pain in the side around waist level or in the lower back
- Nausea and vomiting
- Mental confusion (more common in the elderly)

### Diagnosis

Diagnosis of a urinary tract infection is based on the patient's symptoms and the results of a urine test. Urine is usually obtained via a clean-catch midstream urine specimen: after cleansing the area around the urethral opening and urinating a small amount into the toilet, the patient then urinates into a small cup or container. If the patient is a small child or an adult who cannot urinate, the doctor may obtain a sample by inserting a catheter into the bladder through the urethra.

The doctor can perform a simple urine test to determine whether an infection is present by looking for an increased number of white blood cells in the urine or for the bacteria themselves. To determine which specific bacterium is causing the infection, the doctor can send the urine sample to a laboratory for culture.

The doctor may order imaging tests if there is evidence that the infection is related to structural abnormalities in the patient's urinary tract. Imaging tests are most commonly ordered for patients who have recurrent UTIs (three or more in a year); young children with UTIs, particularly boys; and patients of any age who have blood in the urine.

# Treatment

Treatment of an uncomplicated urinary tract infection in most women is a three- to seven-day course of an antibiotic given by mouth. Adult males usually require two weeks of antibiotic treatment, and children are usually given a ten-day course of antibiotics. The doctor may also prescribe a medication that numbs the pain of urination while the infection is being treated. These drugs may discolor the urine either blue or bright orange.

If the patient has pyelonephritis, intravenous antibiotics may be needed, along with intravenous fluids if the patient is vomiting and cannot keep anything in the stomach. The patient will usually be given oral antibiotics for ten to fourteen days after the intravenous antibiotics are stopped. A patient who is very sick with pyelonephritis, is pregnant, or has kidney stones may be admitted to the hospital for treatment.

The doctor may recommend surgery for children or adults with anatomical abnormalities of the urinary tract in order to reduce the risk of recurrent UTIs and permanent damage to the kidneys.

### Prognosis

Most people with uncomplicated cystitis recover in a few days without any long-term complications. About 1 percent of patients with pyelonephritis die following the spread of the infection from the kidneys to the bloodstream. Those most at risk of death from a severe urinary tract infection are the elderly, people with sickle cell anemia, people with cancer, and people with chronic kidney disease.

# Prevention

There are several precautions to reduce their risk of urinary tract infections:

- Drink plenty of water and other fluids. There is some evidence that cranberry juice is particularly beneficial because it prevents bacteria from attaching to the cells of the tissues lining the bladder.
- Empty the bladder regularly, particularly after sexual intercourse.
- Women using the diaphragm as a method of birth control may wish to consider another method.
- Cleanse the genital area from front to back after using the toilet. This helps to prevent bacteria from the area around the anus from getting into the urethra.
- Urinate promptly when the bladder begins to feel full rather than holding in the urine for long periods of time. In addition, empty the bladder as completely as possible.
- Avoid the use of deodorant sprays or other feminine hygiene products that can irritate the urethral opening; change tampons or napkins frequently during menstrual periods.
- Wear underwear made of cotton rather than nylon or other synthetic fabrics.

In some cases the doctor may prescribe a low-dose antibiotic to be taken after sexual intercourse to prevent a urinary tract infection.

# The Future

UTIs are likely to continue to be commonplace illnesses in the general population. Current research includes studies of electrochemical chips and other new technologies for rapid identification of the bacteria that cause UTIs.

SEE ALSO Diabetes

# For more information

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# WORDS TO KNOW

- **Bladder:** A hollow organ in the lower abdomen that collects urine from the kidneys and stores it prior to urination.
- **Catheter:** A thin tube inserted into the urethra to drain urine from the bladder.
- **Cystitis:** The medical term for an infection of the urinary bladder.
- **Pyelonephritis:** The medical term for a urinary tract infection that has spread from the bladder

or other parts of the urinary tract upward to the kidneys.

- **Ureter:** A muscular tube that carries urine from the kidney to the bladder.
- **Urethra:** The thin tube that connects the bladder to the outside of the body.
- **Urologist:** A doctor who specializes in diagnosing and treating disorders of the kidneys and urinary tract.
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# V

Genetic

Infection

Multiple

Other

Unknown

Injury

# m

# Vision Disorders

Vision disorders are a group of diseases and disorders that affect the sense of sight. Some are caused by infections or allergies, like conjunctivitis, but most are caused by abnormalities in the structure of the eye or changes in the eye over time as a result of aging. Some diseases that affect the whole body, like diabetes, may eventually cause vision problems in addition to other symptoms.

Refractive errors are a type of vision disorder in which the person cannot see clearly because the eye cannot properly focus the light that enters the eye. Hyperopia (farsightedness), astigmatism, and myopia (nearsightedness) are types of refractive error. They develop when the lens of the eye is too strong or too weak for the length of the person's eyeball or when the cornea (the transparent front part of the eye) has an abnormal curvature. Strabismus is a vision disorder in which the eyes are not aligned because the muscles that control the movements of the eyes are weak and uncoordinated.

Cataracts and glaucoma are vision disorders associated with the aging process. A cataract is a yellowish or cloudy discoloration of the lens of the eye, while glaucoma can lead to permanent damage to the optic nerve and eventual blindness.

**SEE ALSO** Astigmatism; Cataracts; Conjunctivitis; Glaucoma; Hyperopia; Myopia; Strabismus

# **u** Vitiligo

# Definition

Vitiligo (pronounced vittle-EYE-go) is a skin disorder in which patches of skin—most commonly on parts of the body that are exposed to sunlight—lose their pigmentation. The patches of depigmented tissue may involve the soft tissues that line the nose and mouth.

# Description

Vitiligo is a skin disorder in which patches of skin, soft tissues lining the nose and mouth, and/or hair gradually lose their color. This loss of color takes place when melanin, a dark brown or reddish pigment produced by cells in the skin called melanocytes, is either destroyed or not produced in the first place. The patches of depigmented skin may grow, shrink, or stay the same size. They usually appear first on the parts of the body that are exposed to the sun, such as the hands, feet, face, lips, and arms. They may spread later to the armpits, genitals, groin area, and the soft tissues that line the nose and mouth. In some cases the white patches may eventually cover the patient's entire body.

Doctors classify vitiligo into three types according to the pattern of the skin patches:

- Focal. The areas of depigmented skin are limited to one or a few parts of the body.
- Segmental. The loss of color occurs on only one part of the body.
- Generalized. There are many patches of white skin on many different parts of the body. This is the most common of the three patterns.

# **Demographics**

Vitiligo is thought to affect between 0.5 and 2 percent of people around the world, as many as 65 million adults. There are between 1 and 2 million people in the United States with the disorder.

Vitiligo can start at any age, but roughly half of patients develop it before age twenty and most patients before age forty. It affects men and women equally, although women are more likely to notice the disorder at an earlier age than men. It also affects people of all races equally;

Also Known As Leukoderma

#### Cause

Unknown; possibly a combination of autoimmune and genetic factors

#### Symptoms

Patches of depigmented skin on various parts of the body

#### Duration

Years

#### Vitiligo



Discoloration on the hands of a woman with vitiligo. © ROB / ALAMY.

however, the depigmented areas of skin are more noticeable on people with darker skin.

In addition to humans, vitiligo is found in some animals, particularly certain breeds of dogs and horses.

# **Causes and Symptoms**

The causes of vitiligo are not completely understood. One reason some researchers think that genetic factors may be involved is that about 30 percent of cases run in families. Another theory is that vitiligo is either an autoimmune disorder or caused by one. Still other researchers think that vitiligo may be stress-related, because some people develop their first symptoms of it following a severe case of sunburn or an emotional shock. None of these theories have been proved so far, however.

In addition to the patches of depigmented skin, the symptoms of vitiligo may include:

- Premature graying (before age thirty) of scalp hair, pubic hair, and eyebrows.
- Uveitis. This is an inflammation of the interior of the eye.
- Depigmented areas on the soft tissues lining the mouth and nose.
- Change in color of the retina of the eye.
#### In the Public Eye: A Television Reporter's Story

Lee Thomas, a television anchor in Detroit, first noticed his vitiligo in 1992, when he was 25 and working for a news station in Louisville, Kentucky. After a haircut he noticed a white spot about the size of a guarter on one side of his scalp. Looking closely, he found other patches on the other side of the scalp, one hand, and inside his mouth. He immediately went to a doctor and was told he had vitiligo. He decided not to let the disease interfere with his goals, though his career depends in part on appearance. On camera, he wears makeup to even out the different colors of his facial skin. "I'm a black man turning white on television and people can see it...If you've watched me over the years, you've seen my hands completely change from brown to white."

Thomas no longer wears makeup outside the studio, a decision made partly for the sake of patients with vitiligo who fear rude or unkind reactions from the public. He admits that he himself went through "times when I would not come out of the house. I call it a mental war. It was me saying, 'I don't want to deal with it today.'" Thomas wrote a book about his experiences and credits the disease with strengthening his inner qualities: "Having this disease forces me to focus on what I am: kind, caring, honest. There are [other] people who have diseases that will kill them."

- Itching of the affected skin. This symptom is usually limited to the early stages of the disorder.
- Increased sensitivity to sun exposure.

Although vitiligo is a skin disorder and not a psychiatric condition, many patients do have strong psychological reactions to the impact of the disorder on their appearance, particularly if the face is affected. In some cultures, people with vitiligo are thought to be evil or infected with leprosy and may be rejected by other members of the community. Many people with vitiligo in the United States become depressed or socially isolated because they are afraid of being stared at or teased.

#### Diagnosis

The diagnosis of vitiligo is based on the patient's history, including a family history of vitiligo. The doctor will also ask about autoimmune disorders, recent sunburns or other injuries to the skin, unusual sensitivity to the sun, or a history of diabetes.

In most cases the doctor can diagnose vitiligo by looking at the patient's skin with a Wood lamp, a special device that uses ultraviolet light to diagnose skin infections and other abnormalities. Another diagnostic technique that can be used is the skin biopsy. To perform a biopsy, the doctor removes a small piece of the affected skin to examine under a microscope. If the patient has vitiligo, the skin sample will usually show a complete absence of melanocytes.

#### Treatment

There is no cure for vitiligo. Medical or surgical treatment is not necessary; some people choose to manage the condition with special cosmetics that cover the white areas and even out the person's skin tone. Even patients who choose medical or surgical therapies may wish to use camouflage cosmetics during their treatment, as it takes between six to eighteen months to complete treatment.

No treatment that is presently available for vitiligo will work for all patients. The choice of treatment depends partly on the number, size, and location of the white patches. Medical treatments for vitiligo are aimed at reducing the contrast between the pigmented and depigmented areas of skin. They include:

- Corticosteroids. These medications are applied directly to the depigmented skin; they work by restoring the color to the white areas. They take about three months for the skin to show improvement.
- PUVA (psoralen/ultraviolet A) therapy. In this type of treatment, the patient takes a drug called psoralen either by mouth or by applying it to the skin as a cream. The patient is then exposed to carefully timed ultraviolet light in the doctor's office. Psoralen reacts with ultraviolet light to darken the skin. Major drawbacks of PUVA therapy are its side effects (chiefly sunburn and too much darkening of the skin) and an increased risk of skin cancer.
- Depigmentation. A drug called Benoquin is applied twice a day to the pigmented areas of the body to lighten them to match the areas that are already white. Depigmentation may be the best choice for people who have vitiligo on more than 50 percent of the body. Its chief drawback is that the person's skin will always be unusually sensitive to sunlight afterward. In addition, this form of treatment is irreversible.

Surgical treatments for vitiligo include:

- Skin grafts. The surgeon removes skin from pigmented areas of the patient's body and places it on depigmented areas. This procedure has a number of drawbacks: it is time-consuming, expensive, painful, and carries some risk of scarring and infection.
- Micropigmentation (tattooing). In this procedure, the doctor implants pigment into the skin with a special surgical instrument. It is usually used only in the area around the lips. One drawback is that it is usually difficult to match the patient's natural skin color.
- Autologous melanocyte transplantation. This procedure is still considered experimental. The doctor removes a sample of the patient's normal pigmented skin and places it in a special culture medium to grow melanocytes. After the melanocytes have

#### WORDS TO KNOW

**Melanin:** A brownish or dark reddish pigment that is the primary determinant of skin, hair, and eye color in humans.

**Melanocyte:** A type of skin cell that produces melanin.

**Retina:** The layer of light-sensitive cells at the back of the eyeball.

**Uveitis:** Inflammation of the interior of the eye.

**Wood lamp:** A special lamp that uses ultraviolet light to detect certain types of skin infections and other disorders, including vitiligo.

multiplied, the doctor transplants them into the areas of depigmented skin. As of 2008, this treatment was very expensive and impractical for most people with vitiligo.

Patients with vitiligo are encouraged to join support groups or seek counseling in order to cope with the emotional effects of the disease.

#### Prognosis

It is very difficult to predict the spread of vitiligo on any patient's body or the outcome of any specific type of treatment.

#### Prevention

There is no known way to prevent vitiligo, because the causes of the disorder are not yet understood.

#### The Future

It is not likely that vitiligo will become either more or less common in the general population than it is at present. Research into the disorder is focused on looking for specific genes that may be related to vitiligo and examining the role of stress in triggering the onset of the disorder. Some evidence has been recently found of a link between vitiligo and variants of a gene called FOXD3.

SEE ALSO Stress; Sunburn

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## W

Genetic

Infection

Injury

Other

Multiple

Unknown

#### Also Known As Verrucae

**Cause** Human papillomavirus (HPV)

#### Symptoms

Small rough benign tissue swellings on the hands, feet, or genitals

#### Duration

A few months to several years



### Warts

#### Definition

Warts are small benign (noncancerous) growths of tissue on the surface of the skin caused by the human papillomavirus (HPV). Most are skincolored but some are darker than the surrounding skin. Dermatologists (doctors who specialize in treating skin disorders) classify warts into four main types according to their location on the body:

- Common warts: Usually found on the fingers, fingernails, and backs of the hands.
- Plantar warts: Found on the soles of the feet. Plantar warts that form in groups are sometimes called mosaic warts.
- Flat warts: Located on the face or (in men) the beard area.
- Genital warts: Found on the external genital areas or around the anus.

#### Description

Warts are small growths on the surface of the skin that result from one or more strains of human papillomavirus infecting the skin through small breaks in the outer surface of the skin. The virus can be transmitted from one person to another; genital warts are highly contagious, whereas the risk of getting common, plantar, or flat warts directly from another person is relatively small.



Warts on a thumb. © MEDICAL-ON-LINE / ALAMY.

The external appearance of warts varies somewhat according to location. Common warts are usually the same color as the surrounding skin and may occur around the edges of the nails where the person has chewed on or cut off a hangnail. The blood vessels inside the wart may look like black dots or seeds.

Plantar warts are usually flat because of the pressure of walking or running. They usually are grayish-yellow in color and may also have black dots inside them like common warts. Plantar warts typically form on parts of the foot that are under pressure from shoes, like the heel or ball of the foot. These warts can be painful as well as unattractive.

Flat warts are smaller (about one-quarter inch [0.64 centimeter]) and smoother than common warts. They are most likely to occur on parts of the body where the skin may be cut or broken by shaving, like the face in men or the legs in women.

Genital warts are a sexually transmitted infection. They usually appear in clusters, which may be very small or may develop into large masses of tissue around the anus or the exterior

genitals. In women, genital warts can grow inside the vagina as well as outside it. Genital warts are not cancerous.

#### **Demographics**

Warts are widespread in the general American population. Between 7 and 10 percent of people develop plantar warts. Flat warts and plantar warts are more common in children and teenagers than adults. Women are slightly more likely than men to develop plantar warts.

#### **Causes and Symptoms**

Warts are caused by infection with the human papillomavirus. There are over 130 strains of this virus that have been identified as of the early 2000s. The virus thrives in warm, moist environments like shower stalls or locker rooms, which is why it can be transmitted indirectly from one person to another. It takes between one and eight months for a wart to appear after the virus has entered a person's body through a break in the skin.

Some people are more susceptible to warts than others, most likely because of genetic factors. No specific genes, however, have been associated with warts.

The symptoms of warts have already been described. Plantar warts are the only type of wart that are usually painful.

#### Diagnosis

Warts can usually be diagnosed by looking at them. When in doubt, a person should consult their doctor to make sure the growth of tissue is really a wart. The doctor can diagnose a wart by scraping off some of the tissue and sending the sample to a laboratory for analysis. Another

## When to Call the Doctor about Warts

A person should see their doctor about warts if they notice any of the following:

- The wart is bleeding heavily or has such signs of infection as red streaks, pus, or a discharge.
- The patient develops a fever of 101°F (38.3°C) or higher.
- The wart is painful.
- The wart is in the anal or genital area.
- The wart has changed its color or overall appearance.
- The patient has diabetes, HIV infection, or an immune system weakened by cancer chemotherapy.

technique, which involves injecting a local anesthetic beforehand, is called a punch biopsy. The doctor takes an instrument called a punch and removes a disk-shaped piece of tissue to send to the laboratory.

#### Treatment

Common or flat warts do not always need treatment. About 30 percent of warts will disappear by themselves within six months. Most will disappear without any treatment within three years. Since warts are not cancerous, most people choose to treat them either because they are unattractive or because (like plantar warts) they cause discomfort when walking.

People can treat common or flat warts at home in several different ways:

- Using over-the-counter preparations containing salicylic acid. It comes in two forms, a liquid to be painted directly on the wart or a sticky plaster to be cut and pasted on the wart. Salicylic acid should not be applied to normal tissue surrounding the wart. It can take as long as twelve weeks to remove warts by this method.
- Covering the wart with adhesive tape or duct tape. This technique works because it forms an airtight environment. The tape is left on

for a week, then removed, and the wart is scraped with an emery board or pumice stone.

- If home treatment does not work, it is best to see the doctor, because the skin growth may be a mole or something other than a wart.
- Genital warts should *never* be treated with home remedies. These warts require professional medical care.

Doctors can treat warts with various medical or surgical techniques:

- Cryotherapy. Cryotherapy uses liquid nitrogen to freeze the warts. The dead tissue in the wart falls away from the skin beneath in about a week.
- Imiquimod. Imiquimod is a topical cream that gets rid of warts by stimulating the body's immune system to fight the virus that causes the warts.
- Cantharidin. Cantharidin is a substance derived from blister beetles. It is a liquid that the doctor applies directly to the skin and covers with a bandage. The cantharidin causes the skin to blister, which lifts the wart from the skin surface and allows the doctor to remove the wart surgically.
- Surgery. The doctor can remove the wart by treating it with an electric needle, then scraping the tissue with a sharp instrument called a curette. Lasers can also be used to remove warts. Both types of surgery can leave scars, however, and are usually used only for warts that have not responded to other types of treatment.

#### Prognosis

As has already been mentioned, about a third of warts will go away without treatment in a few months. Warts often regrow after the skin has healed because the virus that causes them remains in the body; plantar warts are especially likely to reappear. Salicylic acid is about 75 percent effective in removing warts even though it is a slow method.

#### Prevention

People can lower their risk of getting or spreading warts by:

• Avoiding cutting, shaving, brushing, or picking at warts. These actions can spread the virus that causes warts into other areas of nearby skin.

#### WORDS TO KNOW

Benign: Not cancerous.

**Cryotherapy:** The use of liquid nitrogen or other forms of extreme cold to treat a skin disorder.

**Plantar:** Located on or referring to the sole of the foot.

**Topical:** Referring to a type of medication applied directly to the skin or outside of the body.

Verruca (plural, verrucae): The medical term for a wart.

- Avoiding biting their fingernails. The virus that causes common warts often enters the skin of the hands through bitten areas of skin.
- Wearing shower shoes in locker rooms or public showers.
- Washing the hands carefully after touching a wart.
- Keeping the hands and feet as dry as possible, since the HPV virus grows and spreads in warm, moist environments.
- Avoiding sexual contact with anyone known to have genital warts. There is also a vaccine for women that prevents infection with the strains of HPV that cause cervical cancer.

#### The Future

Common, flat, and plantar warts are widespread minor health problems that are likely to continue to affect children as well as adults. It is possible that researchers may eventually identify specific genes that make some people more susceptible to getting warts.

**SEE ALSO** HPV infection

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### West Nile Virus Infection

#### Definition

West Nile virus (WNV) infection is a disease spread by mosquitoes. Severe forms of the infection may be called West Nile encephalitis (if brain tissue is inflamed) or West Nile meningitis (if the membranes covering the brain become inflamed).

#### Description

West Nile virus infection is caused by a virus related to those that cause yellow fever and dengue. It can be classified as a zoonosis because it is a disease that can be spread to humans from animals. It is also called an emerging infectious disease (EID) because it has become more widespread in the last 20 years and is likely to become more common in the future. Most people who become infected with the virus have no noticeable symptoms; about 20 percent develop a mild flu-like illness that lasts about a week; a few people develop a serious illness as a result of WNV causing inflammation of the central nervous system (the brain and spinal cord).

Also Known As

WNV, West Nile fever

#### Cause

Virus from infected birds transmitted by mosquitoes

#### Symptoms

Chills, fever, drowsiness, sweating, headache, weakness, nausea and vomiting

Duration Three to ten days

#### **Demographics**

WNV is increasingly widespread in both tropical and temperate regions of the globe. In North America, epidemics of WNV are most likely to occur in August and September, when mosquitoes are most active. The disease is not a serious infection in 80 percent of people exposed to it; out of 151 people who become infected with the virus, 110 will not notice any symptoms at all; forty will be sick for about a week; and one person will become severely ill. The risk of severe illness in North America is



greatest among people over fifty, pregnant women, people who have recently had cancer chemotherapy, people with diabetes, and people with weakened immune systems. As far as is known, WNV affects all races and both sexes equally. *Magnified image of the West Nile virus.* © SCOTT CAMAZINE / ALAMY.

The West Nile virus is thought to have developed in Uganda about a thousand years ago. It was first identified as a distinct cause of disease in 1937 in a feverish woman in central Africa by researchers who were studying yellow fever. WNV was not described as a cause of severe disease in older adults until an outbreak took place in Israel in 1957. The virus reached North America some time in the 1990s; the first outbreak that attracted public attention occurred in New York, New Jersey, and Connecticut in 1999. The disease has since been reported across the United States. According to the Centers for Disease Control and Prevention (CDC), the total number of deaths in the United States from WNV between 1999 and 2007 is 1,060; most have been in elderly people.

#### **Causes and Symptoms**

West Nile virus infection is caused by a virus transmitted from infected birds to humans through mosquitoes that first bite the birds and then bite people. Crows, ravens, robins, and blue jays are the birds most likely to carry the virus; the CDC reported in 2007 that at least 138 different species of birds in North America have been found to carry WNV, and forty-three different species of mosquitoes. The first major outbreak of West Nile infection in the United States was preceded by the sudden death of large numbers of birds in the New York City area.

#### The Mysterious Death of Alexander the Great

One medical puzzle of ancient history is the death of Alexander the Great (356-323 BCE) at the young age of thirty-two. Alexander returned to Babylon in the spring of 323 BCE after his conquest of India. He fell sick with a fever and died after a two-week illness, which has been variously attributed to typhoid, influenza, liver failure caused by heavy drinking, and poisoning by enemies in his court.

In 2003, however, two researchers in Virginia and Colorado suggested that Alexander might have died from West Nile encephalitis. They noticed an interesting detail recorded by the Greek historian Plutarch: "When [Alexander] arrived before the walls of [Babylon], he saw a large number of ravens flying about and pecking one another, and some of them fell dead in front of him." Epidemics of WNV are often preceded by the death of infected birds, so the American researchers thought that Alexander's symptoms might have been related.

Others disagree, saying that the time course of West Nile encephalitis does not fit what is known of Alexander's last days. The mystery is not likely to yield a final solution, however, as doctors in the ancient world did not use the same terms that are used today to describe symptoms of illness, and translators of ancient Greek documents do not always agree about the meaning of specific words.



Statue of Alexander the Great. SHUTTERSTOCK.

The virus can be transmitted by mosquitoes to bats, horses, dogs, cats, squirrels, skunks, and rabbits as well as to people. The mosquitoes that carry the virus are called vectors, and the infected animals that harbor the virus are called hosts. The virus can also be transmitted by blood transfusions, organ transplants, and from a nursing mother to her baby through breast milk. People cannot get WNV, however, by touching or kissing someone with the virus; there are also no reported cases of people getting West Nile infections from household pets.

When a mosquito carrying WNV bites a human, the virus is transmitted directly into the bloodstream. Once in the bloodstream, WNV multiplies for an incubation period of three to fourteen days. The 20 percent of people who feel symptoms of WNV infection typically have chills, a low-grade fever, headache, sore throat, and nausea and vomiting. About half these patients will also develop a mild rash on the upper chest. In most cases the illness lasts about a week, although the person may feel tired for several weeks afterward.

West Nile encephalitis or meningitis develops when the virus reaches the tissues of the brain or the tissues that cover the brain and the spinal cord and multiplies there. People with this severe form of West Nile infection develop a stiff neck, severe headaches, and feel mentally confused. They have weak or partially paralyzed muscles and may lose consciousness.

#### Diagnosis

The symptoms of WNV are similar to those of other viral infections. A definite diagnosis can be provided by testing blood or spinal fluid for antibodies against the virus. Since 2003, the Food and Drug Administration (FDA) has approved two rapid diagnostic tests for detecting the West Nile virus; one, developed in Australia, gives results in two hours. The other test was developed in Canada and was approved for use in the summer of 2007; it gives results in only fifteen minutes.

#### Treatment

There is no specific treatment for West Nile virus. Most people who develop fever, nausea, or other symptoms recover without seeing a doctor because the symptoms are similar to those of other mild viral infections. The National Institute of Allergy and Infectious Diseases (NIAID) is currently testing a new drug treatment for WNV that was expected to complete its Phase I clinical trial by the end of 2008.

People who develop West Nile encephalitis or West Nile meningitis should be taken to the hospital for specialized treatment to help them breathe. They may also be given intravenous fluids to prevent dehydration and intensive nursing care.

#### Prognosis

Most people who have a mild form of West Nile virus infection recover in about a week with no long-term health problems. Those with severe infections, however, have a 10 percent risk of dying. Those who survive may suffer permanent brain damage or paralysis similar to that caused by polio.

#### Prevention

There is no vaccine against WNV for humans that has completed clinical trials, although a vaccine for horses has been developed and approved for use. The most important precautions that humans can take are controlling mosquitoes and preventing themselves from being bitten by mosquitoes. The CDC recommends the following steps:

- Once a week, drain water from birdbaths, outdoor buckets, flower pots, swimming pool covers, and other water-filled containers where mosquitoes can lay their eggs and breed. Empty children's wading pools when they are not being used.
- Use insect repellent when outdoors; apply it to exposed skin and spray clothing with repellent.
- Wear long-sleeved shirts and long pants when the weather permits.
- Try to stay indoors in early morning and early evening, because mosquitoes are most active at those times of day.
- Keep window and door screens in good repair so that mosquitoes cannot get inside the house.
- Cover baby strollers or carriers with mosquito netting when taking the baby for a walk outdoors.
- Do not handle any dead birds found outside. Contact the local health department first and ask for instructions on reporting and disposing of them.

#### The Future

It is likely that a vaccine for humans and a specific treatment for West Nile virus will be approved by the FDA within the next few years. About

#### WORDS TO KNOW

**Dengue:** A tropical disease caused by a virus similar to the virus that causes West Nile infection. It is also spread by mosquitoes.

**Emerging infectious disease (EID):** A disease that has become more widespread around the world in the last twenty years and is expected to become more common in the future.

Encephalitis: Inflammation of the brain.

- **Host:** An organism that is infected by a virus, bacterium, or parasite. Birds, humans, and other animals may be hosts of West Nile virus.
- **Meningitis:** Inflammation of the membranes that cover the brain and line the brain and spinal cord.
- **Vector:** An animal that carries a disease from one host to another.

450 different chemicals that appear to be effective against viruses have been identified since 2003 as promising enough to justify further testing. There were also two potential vaccines against WNV undergoing clinical trials at the NIAID Vaccine Research Center in Maryland in 2008.

#### SEE ALSO Encephalitis; Meningitis

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# Hiplash

#### Definition

Whiplash is a general term for injuries to the neck caused by stretching of the spine and the soft tissues in the neck when the head is moved sharply forward and then snapped back. It is not a formal medical term; doctors are more likely to use phrases like "cervical sprain" to describe whiplash.

#### Description

Whiplash injuries most commonly occur when a person is in a car or amusement park ride and is struck from behind. The impact pushes the car forward, which in turn throws the occupant's head forward suddenly and then sharply backward, causing overstrain on the muscles or tearing of other soft tissues in the neck and shoulders. Some people feel pain at the time of the accident, while others may not feel the effect of the whiplash injury until several days later. Symptoms may range from soreness or stiffness in the neck and shoulders for a few days to severe pain and disability from injuries to the vertebrae in the neck.

#### Demographics

Whiplash injuries are quite common in the United States and Canada, particularly those related to auto accidents. The National Highway Traffic Safety Administration (NHTSA) estimates that there are between 750,000 and 900,000 whiplash injuries related to automobile travel each year in the United States; about 40 percent of these are caused by rearimpact collisions. They are among the most frequently reported injuries in insurance claims, amounting to about \$8.5 billion in insurance payouts each year.

#### Also Known As

Whiplash-associated disorders, WAD, hyperextension injury, cervical sprain

#### Cause

Rear-end collisions, falls, sports injuries, criminal assault

#### Symptoms

Neck soreness, headaches, shoulder pain, ringing in the ears, dizziness, tiredness

#### Duration

A few days or weeks

#### Whiplash

Illustration of how the brain is forced against the back and front of the skull during whiplash. © NUCLEUS MEDICAL ART, INC. / ALAMY.



Females tend to be at greater risk of whiplash injury than males regardless of whether they are driving the car or riding as a passenger. Apart from gender, drivers are more likely to get a whiplash injury than a front-seat passenger, possibly because drivers are more likely to move forward to try to control the car when hit from behind whereas passengers are more likely to be leaning further back in their seats. Passengers in the rear seats of a car have a slightly lower risk of whiplash injury than drivers or front-seat passengers.

#### **Causes and Symptoms**

Although most whiplash injuries are caused by rear-end auto collisions, they can also be caused by:

- Amusement park rides, particularly roller coasters and bumper-car rides
- Falling from a horse, bicycle, or motorcycle
- Railroad and airplane accidents
- Contact injuries in sports like football, wrestling, or judo
- Being hit on the head by a falling object
- Being shaken or hit on the head by an abusive parent or other attacker

Doctors do not agree, however, on exactly what causes the pain of whiplash in terms of the structures of the human neck. Some think that whiplash injury is caused by hyperextension of the neck, which is the medical term for stretching the neck beyond its normal range of motion. Other doctors think that the biological cause of whiplash injuries is damage to nerve endings in the neck.

#### Home Treatment for Whiplash

Home care for whiplash includes applying heat or cold to the neck and taking mild over-thecounter pain relievers. A common recommendation is to apply ice to the patient's neck for twenty minutes at a time each hour for the first twenty-four hours while the patient is awake. The ice should not be applied directly to the skin but placed inside an ice pack or wrapped in a towel. After the first day or two of recovery, the patient should use a heating pad or other method of applying heat to warm and loosen the neck muscles for stretching exercises.

Acetaminophen or ibuprofen are recommended instead of aspirin for pain relief and to bring down inflammation in the soft tissues of the neck. If the pain is severe, the doctor may prescribe a pain reliever containing codeine or a muscle relaxant. These medications should be taken at bedtime as they often cause drowsiness. There is no clear relationship between an automobile's speed at the time of an accident and a passenger's risk of a whiplash injury; people have reported whiplash injuries when their car was traveling at no more than 15 miles per hour (24 kilometers per hour).

Some doctors use a four-level scale for assessing the symptoms of a whiplash injury. The scale was first drawn up by a task force in Quebec in 1995.

- Level 1: Soreness or stiffness in the neck but there are no physical signs that can be detected by a doctor.
- Level 2: The patient complains of pain and the doctor finds a decreased range of motion in the neck.
- Level 3: The person has pain moving from the neck into the shoulders and arms plus such symptoms as insomnia, weakness, headache, blurred vision, or ringing in the ears.
- Level 4: In addition to the patient's symptoms, the doctor can detect dis-

locations or fractures of the bones in the neck, or injury to the spinal cord.

#### Diagnosis

People with only mild whiplash injuries may simply treat themselves at home (see sidebar). While it is not always necessary to see a doctor for whiplash, people who notice tingling or numbness in their arms or legs, loss of function in their limbs, severe pain when trying to move the neck, pain moving from the neck into the shoulders, headache, or dizziness should see their doctor or go to a hospital emergency department as soon as possible.

Doctors diagnose the seriousness of whiplash injuries by gently moving the patient's head to see how far the neck can move in different directions. They will also press on different parts of the neck to see whether there are any localized sore spots. X rays and other imaging studies may be ordered to check for bone fractures or other signs of serious injury to soft tissues.

#### Treatment

Treatment for mild whiplash injuries may involve massage therapy and heat application in addition to treatment at home with bed rest, ice, and mild pain relievers. Within three to four days of the injury, the patient will be asked to start range-of-motion exercises to keep the neck flexible. These exercises usually involve gently rolling the head from side to side or bending the neck backward and forward. If the pain persists for several months, the doctor may recommend physical therapy to strengthen the neck muscles. The doctor may also inject a local anesthetic to reduce muscle spasms before the patient begins the range-of-motion exercises.

Cervical (neck) collars are not used as often now as they were some years ago. The reason for this change is that wearing a collar for several weeks or months causes the muscles in the neck to lose their strength. People who have trouble sleeping because of whiplash pain, however, may be given a cervical collar to wear while they are sleeping.

Some people are helped by alternative and complementary treatments for whiplash injuries. The approaches that have been found helpful include acupuncture, massage therapy, and chiropractic.

#### Prognosis

Most people feel better within a few days or weeks after a whiplash injury. Severe damage to the bones in the neck or to the spinal cord, however, can result in permanent disability and chronic pain. There is a 40 percent chance of having some symptoms as long as three months after a whiplash injury, and an 18 percent chance that the symptoms will last for as long as two years, but there is no reliable way to predict a specific individual's risk of falling into one of these two groups.

#### Prevention

Head restraints have been mandated in new passenger cars manufactured in the United States since 1969. The effectiveness of these restraints in preventing whiplash injuries is debated by specialists who study the effects of whiplash injuries on the human body, however. One problem is that people do not always adjust their head restraint to the proper height for the size of their head and neck. The head restraint should be

#### WORDS TO KNOW

**Acupuncture:** A form of alternative medicine in which very fine needles are inserted into the skin at specific points on the body for pain relief.

**Chiropractic:** A form of alternative medicine that treats disorders of the joints and

muscles by adjusting the patient's spine or other joints.

**Hyperextension:** Stretching or moving a part of the body beyond its normal range of motion.

adjusted so that the middle of the headrest is even with the upper tips of the ears.

Another problem is that the stiffness of a car seat affects the likelihood of a whiplash injury as much as the quality of the head restraint and its proper adjustment. Safety engineers are still working on designing better head restraints, including some that adjust their position automatically when the driver moves the seat forward or backward.

#### The Future

Some causes of whiplash injury, such as abusive or criminal attacks or accidents involving public transportation, would be difficult to prevent entirely. The risk of sports injuries to the neck can be lowered somewhat by redesigning shoulder pads and other protective equipment and by teaching athletes to minimize their risk of neck injuries. Better automobile design might help also, but even the best-designed seat or head restraint will not be effective if drivers and passengers fail to wear seat belts and adjust their head restraints correctly.

**SEE ALSO** Shaken baby syndrome

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## Whooping Cough

#### Definition

Whooping cough is a highly contagious and potentially fatal bacterial infection of the upper respiratory tract.

#### Description

Whooping cough, or pertussis, is a potentially severe upper respiratory infection characterized by spells of intense coughing that end in a whooping sound when the person is finally able to catch their breath. It can affect people in any age group, but is most common (and most serious) in infants and young children. The organism that causes whooping cough affects only humans; it is not spread by animals.

The early symptoms of whooping cough resemble those of the common cold—runny nose, sneezing, general unwell feeling—and the disease is often mistaken for an ordinary cold. After about a week of cold-like symptoms, however, the patient develops episodes of severe coughing that can bring up thick phlegm (mucus) from the throat. The coughing may be intense and spasmotic enough to cause vomiting or cause the patient to turn red or blue in the face. At the end of the coughing attack, the patient may make a whooping or crowing sound as they gasp to take in their next breath of air.

#### Demographics

At one time whooping cough was a leading cause of death in infants and toddlers, causing 3,000 to 5,000 deaths in the United States Also Known As Pertussis

**Cause** A bacterium

#### Symptoms

Runny nose, sneezing, fever, coughing episodes ending in a whooping sound

Duration

Three to six weeks

#### Whooping Cough

Image of whooping cough bacteria. CNRI / PHOTO RESEARCHERS, INC.



every year. Even though vaccines against whooping cough have been available since the 1940s, the disease is still one of the leading causes of vaccine-preventable deaths worldwide. The World Health Organization (WHO) estimates that there are between 30 and 50 million cases of pertussis each year around the world, and 300,000 deaths. Ninety percent of cases of whooping cough occur in the developing world. Widespread vaccination against the disease, however, has lowered the death rate in the United States to fewer than thirty cases per year.

In the United States, cases of whooping cough tend to cluster in cycles, with peaks every three to four years. Outbreaks of whooping cough are seasonal, with most cases occurring between June and September. On average there are about 2.7 cases of whooping cough per 100,000 in the general population in North America.

Most people who get whooping cough in the United States are unimmunized children or older teenagers and adults whose full immunity has faded. The classic symptoms of whooping cough are not often seen in this latter group, who are likely to have cough for more than three weeks. According to the Centers for Disease Control and Prevention (CDC), 29 percent of patients with whooping cough are younger than one year; 12 percent are aged one to four years; 10 percent are aged five to nine years; 29 percent are aged ten to nineteen years; and 20 percent are older than twenty years.

As far as is known, males and females are equally affected by the disease. Caucasians appear to be more likely to get whooping cough than either African Americans or Native Americans.

#### **Causes and Symptoms**

Whooping cough is caused by a bacterium known as *Bordetella pertussis*, an organism that appears to live only in humans. The organism is spread primarily by droplets in the coughing of infected individuals. When someone breathes in some of these droplets, the bacterium attaches itself to the tissues that line the throat and upper respiratory tract and multiplies. The patient usually begins to feel sick within three to twelve days after being infected.

The symptoms of whooping cough depend on the stage of the illness:

- Early phase (lasts one to two weeks). The patient appears to have an ordinary cold, with runny nose, sneezing, and nasal congestion. There may be low-grade fever and runny eyes. The disease is most likely to be spread to others at this stage.
- Coughing stage (one to two weeks). The patient has spells of intense coughing that may last for several minutes and end

in a whooping sound as the child struggles for breath. Infants younger than six months do not usually make the whooping sound but may become completely exhausted. The child may turn red in the face and vomit at the end of the coughing spell. Adults with whooping cough may get headaches during this stage. The coughing is usually worse at night. In some cases the patient develops pinpoint-sized red marks in the upper chest or the whites of the eyes caused by the breaking of tiny blood

#### **Opposition to Vaccination**

Since the early 1990s public health doctors have become increasingly concerned about a growing trend among parents in some parts of the United States to refuse to have their children vaccinated against whooping cough and other childhood diseases. Most of these parents are too young to remember when such diseases as smallpox, polio, diphtheria, and whooping cough were real threats to children's lives, but because they have heard rumors that vaccines can cause such disorders as autism and multiple sclerosis, they choose not to have their children vaccinated. They think that the risks of side effects from the vaccine are greater than the risks that their children will get a potentially lifethreatening disease.

The problem is that when the rate of vaccination in a community drops below a certain point, diseases that were once rare begin to make a comeback. In parts of Colorado the rate of whooping cough has nearly doubled since the mid-1990s. Over the long term, the children who are not vaccinated are likely to come down with a serious illness themselves as well as putting others in their communities at risk. It may take a major outbreak of whooping cough or another vaccine-preventable disease to change some parents' minds about the benefits of vaccination. vessels during the coughing spells. These little marks are called petechiae.

• Recovery stage (one to two weeks). The child begins to feel better but continues to cough occasionally.

#### Diagnosis

In most cases the doctor will make the diagnosis on the basis of the patient's physical symptoms, a history of exposure to others with whooping cough, and the patient's record of immunization against whooping cough. The doctor may take a blood test to see whether the patient has a higher than normal number of lymphocyte white blood cells or order a chest x ray to see whether the patient has developed pneumonia, but neither of these tests is specific for whooping cough. To make the diagnosis definite, the doctor can take a sample of fluid from the patient's nose or throat on a cotton swab and send it to a laboratory for analysis.

#### Treatment

Treatment of whooping cough depends partly on the patient's age and partly on the severity of the disease. Infants younger than six months often need hospitalization so that they can be given oxygen, have mucus removed from their airway, and fed intravenously if necessary. Very young infants are at greatest risk of ear infections, seizures, or other complications of whooping cough.

Older children, teenagers, and adults should stay home from school or work in order not to give the disease to other people. They should rest in bed if at all possible. In most cases the doctor will prescribe an antibiotic medication, usually for two or three weeks. Antibiotics can shorten the duration of the illness and also shorten the length of time that the patient is contagious. In some cases the doctor will prescribe antibiotics for other members of the patient's family to reduce their risk of getting the disease.

Over-the counter cough medicines are not usually very helpful in relieving the sore throat and coughing spells of whooping cough. A coolmist vaporizer and drinking lots of fluids are usually more effective.

#### Prognosis

Older children, adolescents, and adults usually recover from whooping cough in five to six weeks without any lasting effects. Infants who have not been immunized against whooping cough and older adults with heart problems or lung disease are at greatest risk of developing pneumonia or other complications of whooping cough. About two-thirds of infants who develop whooping cough will need hospital treatment; the average length of stay for a baby with whooping cough in 2004 was seven days. Of the deaths caused by whooping cough in the United States in the early 2000s, 99 percent were in infants.

#### Prevention

The best protection against whooping cough is immunization. Since the 1940s, the vaccine that protects against pertussis has been combined with vaccines against diphtheria and tetanus in a single vaccine. The American Academy of Pediatrics (AAP) recommends a total of five doses of the combined vaccine in children between the ages of two months and six years. Neither the vaccine nor getting the disease confers permanent immunity against whooping cough, however; the effectiveness of the vaccine fades away in three to five years after the last shot.

Because an increasing number of cases of whooping cough are being diagnosed in teenagers, in 2005 the AAP recommended that all teenagers receive an additional booster shot of vaccine. This measure is intended to protect younger children as well as the adolescents, because the evidence indicates that many cases of whooping cough in infants are caused by the infection being transmitted to them by older family members. For adults less than sxity-five years of age, one booster of whooping cough vaccine should be given combined with the once every ten years tetanus/ diphtheria booster. This is especially important in prospective parents and grandparents and those health care workers who work in pediatrics.

#### The Future

Whooping cough is likely to be a major public health problem even in developed countries for the foreseeable future because of some people's opposition to vaccination. In the 1970s there were a number of lawsuits filed by people who felt that they had been injured by the diphtheria/ tetanus/pertussis vaccine. While it was not clear in many of these cases that the person's health problems were caused by the vaccine, Congress passed a law in 1986 that requires doctors to give a child's parents an information sheet about a vaccine before giving the child the shot. The doctor is also required to report any side effects of the vaccination to a national reporting system.

#### WORDS TO KNOW

**Pertussis:** The medical name for whooping cough.

**Petechiae:** Tiny reddish or purplish spots in the skin caused by the breaking of small

blood vessels during intense coughing or vomiting.

**Phlegm:** Thick mucus secreted in the throat and lungs during an upper respiratory infection.

Vaccines licensed for use in the United States must go through a three-stage process of clinical trials that often takes ten years or even longer before the new vaccine is approved. A newer form of the diphtheria/tetanus/pertussis vaccine was developed in 1991 and replaced the older vaccine completely by 2002. It is reported to have lowered the rate of side effects from vaccination in the United States by 90 percent.

SEE ALSO Common cold; Tetanus

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## Х

#### Genetic

Infection

Injury

Multiple

Other

Unknown

#### Also Known As XP

**Cause** Defects in several genes

#### Symptoms

Sunburn after minimal sun exposure; dry, thin skin; discolored patches of skin; eye problems

Duration

Lifelong



## Xeroderma Pigmentosum

#### Definition

Xeroderma pigmentosum, or XP, is a rare condition in which skin cells are not able to repair damage caused by exposure to the ultraviolet portion of sunlight. It was first described in 1874 by two Austrian dermatologists, Ferdinand von Hebra (1816–1880) and Moritz Kaposi (1837–1902). Dr. Kaposi gave the disorder its name, which means "dry discolored skin," in 1882. XP was not understood to be caused by genetic mutations until 1968, however.

#### Description

Xeroderma pigmentosum is a rare skin disorder caused by mutations in any of eight different genes that govern the ability of skin cells to repair damage to DNA (the genetic material inside a cell) caused by exposure to sunlight. In a normal person, exposure to the ultraviolet radiation in sunlight causes damage to the DNA of the cells in the upper layer of the skin. This damage is fixed by a mechanism known as nucleotide excision repair, or NER. In patients with XP, however, the enzymes that carry out NER are either reduced in effectiveness or missing entirely. When the damage to the cell's DNA cannot be repaired, the DNA itself may mutate, leading to the death of skin cells or to skin cancer. The average child with XP develops the first signs of skin cancer around age eight.

#### Xeroderma Pigmentosum

This student with xeroderma pigmentosum has gym classes in the auditorium of her local school because she cannot be exposed to sunlight. AP IMAGES.



Xeroderma pigmentosa progresses in three stages:

• Stage 1. The child's skin is healthy at birth but around six months of age the skin develops reddish, scaly patches with heavy freckling or sunburn after even short exposure to sunlight. At first only the

face and other areas exposed to direct sun are affected, but gradually the skin changes appear on the neck, lower legs, and even the chest.

- Stage 2. The skin develops irregular patches of lightened or darkened skin with spidery patterns of blood vessels appearing underneath. The skin also becomes much thinner.
- Stage 3. This stage begins when the child is four to five years old. Skin cancers and scaly patches known as solar keratoses appear.

About 80 percent of patients with XP develop eye problems related to sun exposure. The eyes may become bloodshot and painfully sensitive to light. Both cancerous and noncancerous growths may appear near the eyes. In a few cases the eyelids may shrink or even disappear entirely.

Between 20 and 30 percent of patients with XP develop symptoms affecting the nervous system in late childhood or early adolescence. These symptoms range from the loss of tendon reflexes and poor coordination to hearing loss and mental retardation. The child's growth may also slow down. These neurologic symptoms tend to get worse over time.

#### Demographics

The frequency of xeroderma pigmentosum varies from country to country. In the United

States, it is estimated to affect between one person in 250,000 and one in 1,000,000. In Japan, however, XP is much more common, striking one person in every 22,000. It is also more common in the Middle East and North Africa than in Europe or North America. The reason for these geographical differences is not yet known for certain but is thought to be associated with the high rates of marriages within extended families in some of these countries.

#### A Very Special Summer Camp

In 1996 the parents of a child with xeroderma pigmentosum had the idea of creating a nighttime camp for patients with XP that would allow them to enjoy the typical activities of a summer camp experience but without the dangers of exposure to sunlight. By 2005 the couple was able to open a permanent retreat house for Camp Sundown, as they named it, in Craryville, New York. The camp's sessions in recent years include a retreat for family members combined with a medical conference that brings researchers from around the world to the camp to share new information with the families and to offer support for living with the limitations that the disorder imposes on family members. The campers themselves enjoy outdoor sports under artificial lights after dark, including an adventure course.

Camp Sundown provides lodging, meals, and supplies to each camper and one family member completely free of charge. Other family members are charged only nominal fees for staying at the camp. In 2007 the camp ran six sessions through the summer and into October, including one week just for teens. Any person or organization can sponsor a camper for one full session with a donation of \$350. That sum offsets the cost of the camper's food, utilities, activities, insurance and transportation. In all countries, XP affects men and women equally. Within the United States, it affects all races and ethnic groups equally.

#### **Causes and Symptoms**

The cause of xeroderma pigmentosum is a mutation in any of eight genes (on eight different chromosomes) known to affect the production of the enzymes responsible for the repair of DNA in sunlight-damaged skin cells. There are eight subtypes of XP that vary somewhat in the severity of symptoms depending on which of the eight genes has been affected by a mutation. The most common subtype, known as XPA, is also the type most likely to be associated with neurologic symptoms.

A child must inherit a defective gene from both parents in order to develop XP. A person who has only one copy of the gene mutation is called a carrier. When both parents are carriers of a recessive gene mutation, there is a 25 percent chance that a child will inherit two mutations and develop XP.

In addition to the symptoms that have already been described, children with xeroderma pigmentosum may also have:

- · Raw patches of broken skin oozing tissue fluid
- Premature aging of the lips, eyes, mouth, and tongue
- Cancer developing on the tip of the tongue
- Limited growth of body hair on the chest and legs
- · Increased vulnerability to infectious diseases

#### Diagnosis

The diagnosis of xeroderma pigmentosa is usually made during the baby's first or second year of life, often following a history of severe and long-lasting sunburn following only brief exposure to sunlight, or heavy freckling on the baby's face. There is usually no family history of the disorder. The doctor will, however, ask the parents about a family history of intermarriage among relatives in the extended families on both sides, as kinship marriages are a risk factor for genetic disorders.

The doctor will examine the baby for signs of eye problems, including clouding of the cornea (the transparent covering of the front of the eyeball that admits light) and skin tumors growing near the eyes. The baby's reflexes and coordination may also be tested. The diagnosis of XP can be confirmed by genetic testing or by tests on skin cells that evaluate whether the cells are unusually sensitive to ultraviolet light. These tests can be performed only in highly specialized laboratories, however.

XP can be diagnosed before birth by amniocentesis or by chorionic villus sampling (CVS). CVS is a test in which the doctor takes a small sample of tissue from the placenta, the organ that forms inside the uterus during pregnancy and links the mother's blood supply with the baby's blood supply. The tissue sample can then be analyzed for evidence of a genetic disorder.

#### Treatment

There is no cure for xeroderma pigmentosum. In addition, any damage that occurs to the skin from sun exposure is permanent and irreversible. The main goals of treatment are therefore to protect the patient from sun exposure and monitor the development of symptoms affecting the eyes and nervous system.

Avoiding sun exposure includes:

- Wearing protective clothing (long-sleeved shirts and full-length pants, shirts with collars, tightly-woven fabrics that don't let light through); wide-brimmed hats; and eyewear specifically made to screen out ultraviolet rays.
- Applying sunscreens of SPF 30 or higher to all exposed areas of skin.
- Avoiding going outdoors during daylight hours as much as possible.
- Limiting outdoor activities to nighttime only.
- Avoiding the use of halogen or fluorescent bulbs indoors; these give off enough ultraviolet radiation to affect people with XP.
- Applying special film that blocks ultraviolet rays to house and car windows.

Monitoring patients with xeroderma pigmentosum includes:

- Regular visits to a dermatologist—at least every three to five months—to check for changes in the skin or early signs of skin cancer.
- Yearly testing of the patient's nervous system.
- Frequent visits to an ophthalmologist (eye specialist) to check for clouding of the cornea, tumors near the eye, or other problems.

Skin cancers are usually removed by surgery as soon as they appear. Solar keratoses are treated with a cream containing a drug called 5-fluorouracil.

#### Prognosis

The prognosis of xeroderma pigmentosum depends on the subtype that the patient belongs to; however, all patients with XP have over a thousand times greater risk of developing skin cancer than people without the disorder. Those with subtype XPA typically develop skin cancer before they are ten years old. Only 40 percent of people with XP live into their twenties. A small percentage of patients with milder symptoms live into middle age. In general, XP reduces a patient's life expectancy by at least thirty years.

#### Prevention

Parents of a child with XP have a 25 percent chance of having another child with the disorder in later pregnancies. They should consult a genetic counselor before considering having other children. In addition, people who have a relative with XP should consider having a genetic test before starting a family.

#### The Future

Researchers are trying to better understand the gene mutations responsible for XP and to see whether there are any other genes associated with the condition that have not yet been identified. Treatments being investigated in 2008 included the use of a drug called isotretinoin in preventing skin cancer in patients with XP and a lotion called T4N5 liposome lotion in protecting against skin damage caused by ultraviolet radiation.

SEE ALSO Skin cancer; Sunburn

#### For more information

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#### WORDS TO KNOW

**Chorionic villus sampling (CVS):** A prenatal test that involves taking a small sample of the placenta, the organ that forms inside the uterus during pregnancy and supplies the baby with oxygen and nutrients carried by the blood.

**Dermatologist:** A doctor who specializes in diagnosing and treating skin disorders.

**Neurologic:** Pertaining to the nervous system.

- Nucleotide excision repair (NER): A mechanism that allows cells to remove damage caused by ultraviolet light to the cell's DNA.
- **Solar keratoses (singular, keratosis):** Rough scaly patches that appear on sun-damaged skin. They are considered precancerous.
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**m** 0

Multiple

Genetic

Infection

Injury

Other

#### Unknown



### Zoonoses

Zoonoses (singular, zoonosis) are infectious diseases that can be transmitted from wild or domestic animals to humans. Some zoonoses can also be transmitted from humans to other animals. The disease agents that can cause zoonoses include prions (abnormal proteins), bacteria, viruses, fungi, and parasites.

A wide variety of animals can serve as vectors, or carriers, of zoonoses. Wild animals and insects that can spread zoonoses include bats, birds, chimpanzees, fleas, flies, mosquitoes, mice and rats, monkeys, opossums, rabbits, snails, and ticks. Domestic and farm animals that can transmit zoonoses include cats, dogs, cattle, goats, horses, pigs, and sheep. Animals can transmit zoonoses through biting humans (plague, Lyme disease, rabies), leaving droppings that contain the disease organism (hantavirus infection, toxoplasmosis), or harboring the organism in their tissues or hair (anthrax).

Zoonoses range from mild diseases like cat scratch fever and cowpox to highly fatal illnesses like Ebola and Marburg hemorrhagic fevers. Some zoonoses, like rabies, appear as isolated cases in humans, while others, like AIDS and plague, can spread directly from one human to another and cause worldwide epidemics. Some doctors think that many diseases that are now spread directly among humans, like measles, tuberculosis, influenza, and the common cold, may have started out as zoonoses thousands of years ago.
**SEE ALSO** AIDS; Anthrax; Avian influenza; Creutzfeldt-Jakob disease; Ebola and Marburg hemorrhagic fevers; Hantavirus infection; Lyme disease; Plague; Rabies; Toxoplasmosis; West Nile virus infection

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### List of Organizations

#### Academy for Eating Disorders

111 Deer Lake Rd., Ste. 100 Deerfield, IL 60015 Phone: (847) 498-4274 Fax: (847) 480-9282 E-mail: info@aedweb.org Internet: http://www.aedweb.org/

#### American Academy of Child and Adolescent Psychiatry

3615 Wisconsin Ave. NW Washington, DC 20016-3007 Phone: (202) 966-7300 Fax: (202) 966-2891 Internet: http://www.aacap.org/

#### American Academy of Pediatrics

141 Northwest Point Blvd. Elk Grove Village, IL 60007-1098 Phone: (847) 434-4000 Fax: (847) 434-8000 E-mail: kidsdocs@aap.org Internet: http://www.aap.org/

#### Alzheimer's Association

225 N. Michigan Ave., 17th Floor Chicago, IL 60601-7633 Phone: (312) 335-8700 Fax: (866) 699-1246 E-mail: info@alz.org Internet: http://www.alz.org/

#### American Academy of Sleep Medicine

One Westbrook Corporate Center, Ste. 920 Westchester, IL 60154 Phone: (708) 492-0930 Fax: (708) 492-0943 Internet: http://www.aasmnet.org/

#### American Cancer Society

1599 Clifton Rd. NE Atlanta, GA 30329 Phone: (800) ACS-2345 Internet: http://www.cancer.org/

#### American Diabetes Association

1701 N. Beauregard St. Alexandria, VA 22311 Phone: (800) 342-2383 E-mail: AskADA@diabetes.org Internet: http://www.diabetes.org/

#### American Foundation for AIDS Research

120 Wall St., 13th Fl. New York, NY 10005-3908 Phone: (212) 806-1600 Fax: (212) 806-1601 Internet: http://www.amfar.org/

#### American Heart Association

7272 Greenville Ave. Dallas, TX 75231 Phone: (800) 242-8721 Internet: http://www.americanheart.org/

#### American Lung Association

61 Broadway, 6th Fl. New York, NY 10006 Phone: (212) 315-8700 Phone: (800) 586-4872 Internet: http://www.lungusa.org/

#### American Medical Association

515 N. State St. Chicago, IL 60610 Phone: (800) 621-8335 Internet: http://www.ama-assn.org/

#### American Obesity Association

8630 Fenton St., Ste. 814 Silver Spring, MD 20910 Phone: (301) 563-6526 Fax: (301) 563-6595 Internet: http://www.obesity.org/

#### American Parkinson Disease Association

135 Parkinson Ave. Staten Island, NY 10305 Phone: (718) 981-8001 Phone: (800) 223-2732 Fax: (718) 981-4399 E-mail: apda@apdaparkinson.org Internet: http://www.apdaparkinson.org/

#### Arthritis Foundation

PO Box 7669 Atlanta, GA 30357-0669 Phone: (800) 283-7800 Internet: http://www.arthritis.org/

#### Autism Society of America

7910 Woodmont Ave., Ste. 300 Bethesda, MD 20814-3067 Phone: (301) 657-0881 Phone: (800) 328-8476 Internet: http://www.autism-society.org/

# Centers for Disease Control and Prevention (CDC)

1600 Clifton Rd. Atlanta, GA 30333 Phone: (800) 232-4636 Internet: http://www.cdc.gov/

#### **Cystic Fibrosis Foundation**

6931 Arlington Rd. Bethesda, MD 20814 Phone: (301) 951-4422 Phone: (800) 344-4823 Fax: (301) 951-6378 E-mail: info@cff.org Internet: http://www.cff.org/

#### **Epilepsy Foundation**

8301 Professional Place Landover, MD 20785 Phone: (800) 332-1000 Internet: http://www.epilepsyfoundation. org/

#### Huntington's Disease Society of America

505 Eighth Ave., Ste. 902 New York, NY 10018 Phone: (212) 242-1968 Fax: (212) 239-3430 E-mail: hdsainfo@hdsa.org Internet: http://www.hdsa.org/

#### March of Dimes Birth Defects Foundation

1275 Mamaroneck Ave. White Plains, NY 10605 Phone: (914) 997-4488 Internet: http://www.modimes.org/

#### Muscular Dystrophy Association-USA

National Headquarters 3300 E. Sunrise Dr. Tucson, AZ 85718 Phone: (800) 572-1717

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E-mail: mda@mdausa.org Internet: http://www.mdausa.org/

#### National Center for Complementary and Alternative Medicine

9000 Rockville Pike Bethesda, MD 20892 Phone: (301) 519-3153 Phone: (888) 644-6226 Fax: (866) 464-3616 E-mail: info@nccam.nih.gov Internet: http://www.nccam.nih.gov/

#### National Diabetes Information Clearinghouse

1 Information Way Bethesda, MD 20892-3560 Phone: (800) 860-8747 Fax: (703) 738-4929 E-mail: ndic@info.niddk.nih.gov Internet: http://diabetes.niddk.nih.gov/

# National Digestive Diseases Information Clearinghouse

2 Information Way Bethesda, MD 20892-3570 Phone: (800) 891-5389 Fax: (703) 738-4929 E-mail: nddic@info.niddk.nih.gov Internet: http://digestive.niddk.nih.gov/ about/

#### National Down Syndrome Society

666 Broadway, 8th Fl. New York, NY 10012 Phone: (800) 221-4602 E-mail: info@ndss.org Internet: http://www.ndss.org/

#### National Eating Disorders Association

603 Stewart St., Ste. 803 Seattle, WA 98101 Phone: (206) 382-3587 Phone: (800) 931-2237 Fax: (206) 829-8501 E-mail: info@NationalEatingDisorders.org Internet: http://www.nationaleating disorders.org/

#### National Fibromyalgia Association

2121 S. Towne Centre Place, Ste. 300 Anaheim, CA 92806 Phone: (714) 921-0150 Internet: http://www.fmaware.org/

#### National Heart, Lung, and Blood Institute Health Information Center

PO Box 30105 Bethesda, MD 20824-0105 Phone: (301) 592-8573 Fax: (240) 629-3246 E-mail: nhlbiinfo@nhlbi.nih.gov Internet: http://www.nhlbi.nih.gov/

#### National Hemophilia Foundation

116 W. 32nd St., 11 Fl. New York, NY 10001 Phone: (212) 328-3700 Phone: (800) 424-2634 Fax: (212) 328-3777 E-mail: handi@hemophilia.org Internet: http://www.hemophilia.org/

# National Institute of Allergy and Infectious Diseases

6610 Rockledge Dr., MSC 6612 Bethesda, MD 20892-6612 Phone: (301) 496-5717 Phone: (866) 284-4107 Fax: (301) 402-3573 Internet: http://www.niaid.nih.gov/

# National Institute of Diabetes and Digestive and Kidney Diseases

Bldg. 31, Rm. 9A04, 31 Center Dr., MSC 2560 Bethesda, MD 20892-2560 Phone: (301) 496-3583 Internet: http://www.niddk.nih.gov/

#### National Multiple Sclerosis Society

733 Third Ave. New York, NY 10017 Phone: (212) 986-3240 Phone: (800) 344-4867 Internet: http://www.nmss.org/

#### National Osteoporosis Foundation

1232 Twenty-second St. NW Washington, DC 20037-1202 Phone: (202) 223-2226 Phone: (800) 231-4222 Internet: http://www.nof.org/

#### Sickle Cell Disease Association of America

231 E. Baltimore St., Ste. 800

Baltimore, MD 21202 Phone: (410) 528-1555 Phone: (800) 421-8453 Fax: (410) 528-1495 E-mail: scdaa@sicklecelldisease.org Internet: http://www.sicklecelldisease.org/

#### World Health Organization

Avenue Appia 20 Geneva 27, 1211 Switzerland Phone: (011-41-22) 791-2111 Fax: (011-41-22) 791-3111 E-mail: inf@who.int Internet: http://www.who.int

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