

CF WORDS TO KNOW

Glossary (v4)

A

- **Abdomen**
Also called the belly. The part of the body below the diaphragm and above the pelvis where the digestive tract, liver, and kidneys are located.
- **Abdominal Wall**
The tissues between the skin and the abdominal cavity. Includes muscle, fat, and soft tissues.
- **ABG**
Short for arterial blood gas test. Blood is taken from a blood vessel in the wrist called an artery. If a person places his fingers over this artery in the wrist, he can feel his pulse. The ABG test measures how much oxygen, carbon dioxide, and acid are in the blood. See also **PaCO₂**; **PaO₂**.
- **ABPA**
Short for allergic bronchopulmonary aspergillosis. This is a complication that is sometimes seen in CF. In ABPA, the airways overreact when they are exposed to a fungus called *Aspergillus*. This allergic reaction can cause symptoms, such as coughing and wheezing, and can reduce lung function. One test used to check for ABPA is the IgE blood level. Usually anti-inflammatory medicine is needed to treat ABPA. See also **allergy/allergies**; **IgE**.
- **Absorb(ed)/Absorbing/Absorption**
The passing of nutrients into the bloodstream. This occurs mainly in the small intestine after food is digested. Nutrients include proteins, fats, and carbohydrates. Vitamins and minerals are also absorbed. See also **digest(ing)/digestion**; **malabsorption**.
- **Abstinence**
To refrain from an activity. For example, to not have sexual intercourse. See also **sexual intercourse**.
- **acapella[®] Vibratory PEP Therapy System**
A handheld device that is used for airway clearance to help move mucus out of the lungs. A person with CF blows into the device and it loosens mucus by making vibrations in the airways. The person needs to huff and cough between a set of blows. A CF health care team member teaches families how to use the acapella[®]. See also **huff(ing)**.
- **Achromobacter (Alcaligenes) Xylooxidans**
A type of bacterium. In CF, *Achromobacter (Alcaligenes) xylooxidans* can sometimes be found in the sputum and may cause lung infections.
- **Acid Reflux**
Fluid from the stomach that comes back up the esophagus. See also **esophagus**; **gastroesophageal reflux**.
- **Action Plan**
See **CF action plan**.
- **Active Cycle of Breathing (ACB)**
Active cycle of breathing, or ACB, is a way to do airway clearance using breathing and huffing exercises to help move mucus out of the lungs. ACB works best when it is done in postural drainage positions. The child needs to cough after huffs. A CF health care team member teaches families how to do ACB. Usually a person has to be at least 9 years old to do ACB well. See also **airway clearance**; **huff(ing)**; **postural drainage and percussion**.
- **Acute**
A disease suddenly getting worse or more severe, or an illness that only lasts a short time. In CF, a chronic disease, acute infections are common. See also **chronic**.
- **Adenovirus**
A common type of virus that can cause upper and lower respiratory tract infections (from the common cold to pneumonia) in people of all ages. Adenovirus can also cause eye infections and acute gastrointestinal infections (vomiting and diarrhea). Some people can have an infection without symptoms. A person can have an adenovirus infection more than once.
- **Adolescence**
The stage of life when the body is undergoing sexual maturation and rapid growth. There are also many other changes in the body and brain development during this time.
- **Adolescents**
Also called teens or teenagers. Children in adolescence stage of life. See also **adolescence**.

- **Adverse Reaction**

Unwanted side effect resulting from a drug or mixture of drugs. See also **side effects**.

- **Advocate**

A person who speaks in favor of the child and his or her rights.

- **Aerobic**

Requiring oxygen. Aerobic exercises (such as running and swimming) are physical activities that make a person work hard. Aerobic exercise increases the need for oxygen.

- **Aerosol**

Particles (very small drops) of a solid or liquid forming a mist that can be inhaled. Some drugs used to treat CF are in this form. See also **nebulizer; particle**.

- **AFB**

Short for acid-fast bacteria. These bacteria are called acid-fast because of the type of stain used to see them under the microscope. The most well known AFB causes tuberculosis (*Mycobacterium tuberculosis*). Other types of AFB can sometimes cause problems for people with CF. The most common AFB infection in CF is caused by *Mycobacterium abscessus*. These AFB infections tend to progress very slowly and can be hard to diagnose. They need longer antibiotic treatment than is needed for other bacteria.

- **Air Compressor**

A machine that connects to a nebulizer by tubing and creates pressure through airflow to turn liquid medicine into a mist. Children with CF can then inhale the medicine during breathing treatments.

- **Air Sacs**

Another name for alveoli, the grape-like clusters at the end of the airways in the lungs where gas exchange takes place. See also **alveoli**.

- **Airway(s)**

Tubes that bring outside air into the lungs. The lungs have many airways of different sizes. The largest airway is the trachea (windpipe) between the throat and the lungs. It branches into smaller airways called bronchi. These divide into the smallest branches called bronchioles. At the end of the bronchioles are the alveoli (air sacs). The airways are shaped somewhat like a tree that goes

from the trunk to the twigs and ends in the leaves. In CF, abnormal mucus clogs the airways. Infection and inflammation cause airway damage over time. See also **alveoli; bronchi; bronchioles; trachea**.

- **Airway Clearance**

A term for techniques a person can use to loosen abnormal (thick, sticky) mucus so it can be coughed or huffed out of the lungs. CF can cause mucus to build up in the airways. The mucus blocks airflow and is a good place for bacteria to grow. There are different ways of doing airway clearance, such as postural drainage and percussion (PD & P), breathing exercises, using handheld devices such as the Flutter® and acapella®, or wearing a percussive vest. See also **acapella® Vibratory PEP System; Flutter® device; percussive vest; postural drainage and percussion**.

- **Albumin**

A type of protein that is found in the body and can be measured in the blood.

- **Allergens**

The things to which people are allergic. Common allergens are dust, pollens, and molds. These are often called the person's allergic triggers. See also **allergy/allergies**.

- **Allergy/Allergies**

The body's overreaction when exposed to a substance that is an allergen. The overreaction occurs when the body has become sensitive to a certain allergen. Allergic reactions may cause a number of symptoms, such as rash or hives, diarrhea, cough, wheezing, or sinus problems. Symptoms vary from person to person.

- **Alveoli**

Tiny grape-like clusters of air sacs at the ends of the airways, deep in the lungs, where the body gets its fresh oxygen supply. The walls of the alveoli are covered with tiny blood vessels. As blood is pumped through these vessels, oxygen passes from the alveoli into the bloodstream. At the same time, carbon dioxide passes out of the bloodstream into the air sacs. With each breath, a person exhales carbon dioxide and inhales oxygen-rich air. A single air sac is called an "alveolus." In CF, mucus can clog both the airways and the alveoli. See also **airway(s)**.

- **Aminoglycoside Antibiotics**

A type of antibiotic that is used to treat certain bacterial infections. Aminoglycoside antibiotics include tobramycin, gentamicin, and amikacin. This type of antibiotic is often used in CF to treat infection in the lungs. Possible side effects include reduced kidney function and hearing problems. Drug blood levels need to be checked when aminoglycoside antibiotics are given by IV.

- **Amylase**

One of the enzymes produced in the pancreas. Amylase helps break down carbohydrate starches in food in the intestine. Amylase is included in pancreatic enzyme supplements in CF.

- **Anemia**

A low red blood cell count. Can be due to diet problems, blood loss, or diseases that affect how the body makes red blood cells.

- **Antibacterial**

Something that kills bacteria or stops them from growing, such as the bleach solution used to clean equipment or antibacterial hand cleaner.

- **Antibiotics**

Drugs used to fight infections caused by bacteria. Antibiotics kill bacteria or stop them from growing. In CF, antibiotics are used to control infections, such as bronchitis. Antibiotics may be taken by mouth, by injection, or by IV. Some antibiotics may be inhaled.

- **Antibodies**

The proteins that the body makes to help protect itself from disease. Antibodies are made to fight specific bacteria or other substances that are foreign to the body. See also **immunizations**.

- **Anti-inflammatory**

Drugs that turn off or prevent inflammation. These drugs may be inhaled, or given by mouth, by injection, or by IV. Some anti-inflammatory drugs, such as prednisone, are corticosteroids. Other anti-inflammatory medicines, such as ibuprofen or montelukast (Singulair®) do not contain steroids. Inflammation occurs in the airways in CF. See also **inflammation**.

- **Antioxidant**

An enzyme or other substance (such as vitamin E) that can counteract the damaging effect of oxidation in the body's tissues. Oxidation is a process that can cause injury to cells. Oxidation also causes metal to rust and vegetables to rot.

- **Anus**

The hole where stool (the solid waste from digested food) passes out of the body. The anus is part of the digestive system.

- **Aspergillus**

A kind of fungus (mold) that is very common in the environment. Some people develop an allergy to *Aspergillus*. In CF, sometimes a person can have a strong allergic reaction to *Aspergillus* called ABPA (allergic bronchopulmonary aspergillosis). See also **ABPA; allergy/allergies**.

- **Asthma**

A chronic disease in which the airways overreact with exposure to a trigger. It is sometimes called reactive airways disease (or RAD, for short). Symptoms of asthma may be triggered by allergens, exercise, infections, or irritants (such as tobacco smoke). Different people are affected by different triggers. During an asthma problem, the airway muscles tighten. This reaction is called bronchospasm. Inflammation develops in the airways in asthma, which causes swelling and more mucus. All of these reactions can cause the airways to narrow and make it hard to breathe. Common symptoms of asthma are coughing, wheezing, and shortness of breath. Asthma can be treated. Some people with CF have asthma. At times, the airways in CF can have asthma-like reactions and may be treated with asthma medicines. See also **anti-inflammatory; bronchodilator; bronchospasm**.

- **Atelectasis**

Collapsed portion of the lung that does not contain air. This is often caused by a buildup of mucus in the airway that prevents air from getting in. Airway clearance helps to get rid of the mucus.

- **Attending Physician**

The doctor who is in charge of a patient's care in the hospital or clinic.

- **Audiometry**

The name used for hearing tests. In CF, hearing problems can sometimes develop as a side effect of certain medicines. One type of medicine that can cause hearing problems is the aminoglycoside antibiotic. See also **aminoglycoside antibiotic**.

- **Autogenic Drainage**

A type of airway clearance that uses patterns of breathing and huffing to help get mucus out of the lungs. Autogenic drainage can be done in any position that is comfortable; gravity is not used to move the mucus. A CF health care team member teaches families how to do autogenic drainage. Usually a person has to be at least 12 years old to do autogenic drainage well because it requires greater concentration and body awareness than some other types of airway clearance.

- **Autosomal Recessive**

How certain genetic traits or problems are inherited. Problems inherited this way occur only when a person receives two gene mutations for the trait, one from each parent. With only one gene mutation for the trait (from either the mother or father, but not both) the person won't be affected by the disease. CF is an autosomal recessive disease. See also **carrier; gene(s)/gene mutation; genetic**.

- **Axillary Temperature**

A temperature reading under the arm. May be used with infants and young children.

B

- **Bacteria**

Tiny one-celled living organisms that may cause infections. People with CF can get bacterial lung infections. Some common bacteria that cause infection in CF are *Pseudomonas aeruginosa* and *Staphylococcus aureus*. There are some types of bacteria that are helpful and live in the body without causing harm. For example, *Escherichia coli* in the intestine help with digestion. See also **infection(s)**.

- **Barrel Chest**

An enlarged rib cage caused by some lung diseases, including CF. This may also be called “increased AP (anterior-posterior) diameter” of the chest (front to back).

- **Baseline Symptoms**

A person's usual symptoms when stable during a well period. Some people have no symptoms when well. Other people with CF may have some baseline cough or mucus production. Baseline digestion symptoms include how many stools a person has every day. Families who have children with CF should know and record their child's baseline symptoms.

- **Beta-Agonist**

A connecting point on the surface of cells. In the airways, a beta-agonist connects to cells to relax the muscles. This can relieve bronchospasm. See also **bronchospasm**.

- **Bicarbonate**

An electrolyte found in the body. Bicarbonate is important in the balance of acid and base in the body. It forms a type of salt with sodium called sodium bicarbonate, the main ingredient in baking soda. See also **electrolytes; sodium**.

- **Bile**

A fluid that is made by the liver. Extra bile is stored in the gallbladder. Bile is carried to the intestines to help digest food. See also **gallbladder; liver**.

- **Bile Duct**

A tube in the liver that carries bile fluid to the intestines to be used for digestion.

- **Blockage**

When something blocks a passage. In the lungs, mucus blockage prevents air from moving freely in and out of the lungs. In people with CF, a blockage of stool or mucus may also occur in the intestine.

- **Blood Clot/Blood Clotting**

Blood that forms a solid mass called a clot. Blood clots are needed to stop bleeding. Blood clots that form in blood vessels when not needed can cause a blockage. If the blood does not have enough vitamin K, it may not clot properly. People with CF need vitamin K supplements. The liver is also important in making blood clot. Liver disease in CF can cause bleeding problems. See also **blockage; vitamin K**.

- **Blood Glucose**

The main sugar in the blood. A major source of fuel for the body. The sugar in blood comes from the digestion of carbohydrates. See also **carbohydrates; CFRD; diabetes**.

- **Blood Sugar**
See **blood glucose**.
- **Body Mass Index (BMI)**
A number that compares a person's weight and height to estimate the amount of body fat and muscle. The CF Foundation recommends that children with CF have a BMI at or above the 50th percentile for age.
- **Bolus Feeding**
A feeding that is given over a short time. A bolus feeding could be with a bottle or could be given through a feeding tube.
- **Bone Density**
A measure of the amount of minerals, such as calcium, found in the bones. This is a measure of how strong the bones are.
- **Bowel Movement (BM)**
A name for the waste from digested food that passes through the intestines. Other names include stool or poop. In newborn infants, the first bowel movement is called meconium. Sometimes in CF the thick meconium gets stuck and causes a bowel obstruction called meconium ileus. See also **distal intestinal obstruction syndrome (DIOS); meconium ileus**.
- **Bowels**
Another name for the intestines. These are the hollow tubes in the abdomen used for digestion and through which stool passes out of the body.
- **Breathing Treatments**
Another name for respiratory treatments where a person inhales medicine as a mist. In CF, there are a number of medicines that are inhaled into the airways from a breathing treatment.
- **Bronchi**
Airways that branch from the trachea and carry air throughout the lungs.
- **Bronchiectasis**
A weakened condition of the airway walls. In CF, damage from mucus obstruction and infections causes bronchiectasis. The weakened airways do not drain mucus well. Bronchiectasis is a common complication in CF. See also **obstruction**.
- **Bronchioles**
The smallest airways in the lungs that branch off from the bronchi. The bronchioles reach deep into the lungs and end in the air sacs or alveoli. See also **alveoli; bronchi**.
- **Bronchiolitis**
An inflammation of the smallest airways called the bronchioles. Usually this is caused by an infection from a virus. It is seen most often in young infants. Infants with CF can have severe bronchiolitis. Common symptoms are wheezing and difficulty breathing. Viruses that cause bronchiolitis include RSV and influenza. See also **influenza; RSV**.
- **Bronchitis**
An inflammation of the bronchi (airways). Usually this is caused by infection from either bacteria or viruses.
- **Bronchodilator**
A type of medicine that relaxes the airway muscles. This helps open the airways so a person can breathe more easily. These medicines are helpful in treating asthma and bronchospasm. See also **bronchospasm**.
- **Bronchoscopy**
A test that uses a bronchoscope to look at the main airways. A bronchoscope is a tube that has a fiber-optic camera and suction. The tube is placed through the nose into the back of the throat and then is passed through the vocal cords into the airways. The doctor can see and take pictures of the airways. Samples of mucus can be suctioned out of the airway for a culture in the lab. A biopsy (small sample) of the airway lining can be removed for study.
- **Bronchospasm**
The tightening of the muscles around the airways. This causes the airways to narrow making it hard to breathe. Bronchospasm occurs in asthma and sometimes in CF. Common symptoms are coughing, wheezing, and shortness of breath.
- **Burkholderia Cepacia Complex**
A group of bacteria called *B. cepacia* for short. In CF, *B. cepacia* can colonize the lungs and can cause lung infection. *B. cepacia* bacteria are often resistant to many antibiotics and may be spread from person to person. People who have *B. cepacia* in their sputum culture are isolated when they are in the clinic and hospital to prevent these bacteria from spreading to others. See also **colonization/colonize; resistance**.

C

- **Calcium**

A mineral that is an important nutrient for the body. Calcium helps make bones strong and is used in other cells as well.

- **Calorie(s)**

A measure of the amount of energy the body gets from food. People with CF need more calories than people who do not have CF. Extra calories are needed to fight infection. People with CF need to eat more calories (a high-calorie diet) if they do not absorb all the food they eat. See also **high-calorie food additives; malabsorption**.

- **Calorie Dense**

A food that has a lot of calories packed in a small amount.

- **Candida**

A kind of fungus (yeast) that is very common in the environment. *Candida* can cause a throat infection (thrush) or a diaper rash. Some people carry *Candida* in the throat or on the skin without an infection.

- **Carbohydrate(s)**

A type of nutrient found in many foods. Examples are sugars and starches. Simple carbohydrates, such as plain jello, do not require pancreatic enzymes for digestion. Carbohydrates are an important source of energy for the body.

- **Carbon Dioxide**

CO₂ for short. This is a gas that the body makes as it uses oxygen. Carbon dioxide leaves the body when a person breathes out. If a person does not breathe well, carbon dioxide can build up in the blood causing problems. The carbon dioxide level can be measured in the blood.

- **Carrier**

A person who has a gene mutation. A carrier has one gene mutation for the disease and one normal gene. If the disease is a recessive genetic disease, such as CF, carriers will not have the disease, but they may pass the disease gene on to their children. When two CF carriers conceive a child, the baby may get a CF gene from each parent. If so, the child will be born with CF. When the baby gets only one CF gene, he or she will be a carrier like the parent. With two normal genes, the baby is not a carrier. See also **gene(s)/gene mutation; genetic**.

- **Catheter**

A catheter is a hollow tube. There are different types of catheters that are put into the body to deliver or drain fluids, such as an IV catheter, urine catheter, or arterial catheter. Sometimes a catheter in the artery or vein is called a line (art line, IV line, central line). See also **central line**.

- **CBC (Complete Blood Count)**

A test that tells the number of red and white blood cells in the blood. If the red blood cell count is low, a person can be anemic and may need treatment. The types and number of white blood cells can be clues to infection in the body.

- **Cells**

The basic units of all living things. The human body has many different types of cells. CF causes exocrine cells (cells that secrete saliva, sweat, tears, and mucus) in various parts of the body to not work normally.

- **Central Line**

An IV catheter placed in a large vein close to the heart. A catheter is a hollow tube. A central line may be needed to give intravenous (IV) medicines such as antibiotics. A central line is placed in the vein while a person is sedated (asleep after taking a medication). A central line may be temporary or may be placed for longer use. Types of central lines include HICKMAN®, BROVIAC®, and PORT-A-CATH®. See also **catheter; intravenous(ly)**.

- **Cervix**

The opening of the uterus between a female's vagina and uterus. See also **uterus; vagina**.

- **Cesarean Section**

Also called C-section. Delivery of a baby by surgery.

- **CF**

Short for cystic fibrosis. See **cystic fibrosis**.

- **CF Action Plan**

A written treatment plan made with the CF health care team that outlines what parents and their children with CF do at home to manage CF.

- **CF Manager**

Refers to what parents and their children (as they grow older) work toward as they learn the skills Watch and Discover and Think and Act to manage CF. To become good CF managers, parents learn how to: 1) prevent problems; 2) treat complications; and 3) communicate with the CF health care team. As CF managers, parents are central in their child's

care and important members of the CF health care team. Parents help their children with CF learn how to manage the disease, too, so that they can become good CF managers and take more responsibility of their own care. See the CF FEP module: *Becoming a CF Manager*. See also **Think and Act; Watch and Discover**.

- **CF-Related Diabetes (CFRD)**

CFRD is a form of diabetes that can occur in CF. Diabetes is a problem in which a person's blood glucose (a type of sugar) level is too high. In CFRD, two things cause problems with high blood sugar: 1) the pancreas may stop making enough insulin because the insulin cells are damaged from CF and 2) a person can also become "insulin resistant" so that the body's cells don't respond normally to insulin that is made. See also **blood glucose; impaired glucose tolerance; insulin**.

- **CFTR**

Short for cystic fibrosis transmembrane conductance regulator. This protein makes a channel in the cell that allows chloride to go in and out of the cell. The gene defect in CF causes problems with the CFTR. This is the basic cell defect in CF. See also **chloride**.

- **Chest Physical Therapy**

Also called CPT for short. This is an older term for airway clearance that uses physical methods to loosen mucus and help get it out of the lungs. See also **airway clearance**.

- **Child Life Specialist**

A health care professional who is trained to help children and their families cope with disease and the tests and treatments required in the clinic, hospital, and at home. A child life specialist helps children understand what is happening in a simple way according to their age and abilities. This helps children deal with their fears about illness and medical tests.

- **Chloride**

Chloride is part of salt and important to the body's cells. The abnormal CFTR protein in CF affects the movement of chloride through the walls of the cells. In turn, this affects how water and sodium move in and out of the cells. Water and sodium affect how thick the mucus is. The high chloride level in sweat (determined through the sweat test) helps diagnose CF. See also **sodium; sweat test**.

- **Chromosomes**

The parts of cells that carry genes. Chromosomes are found in every cell of the body. Normally a person has 23 pairs of chromosomes (a total of 46 chromosomes). When a child is conceived, each parent provides half the chromosomes, one for each of the 23 pairs. Each chromosome contains hundreds of genes. The CF gene is carried on the chromosome number 7. See also **conceive(d)**.

- **Chronic**

A condition that lasts a long time. Most chronic diseases cannot be cured. Treatments are used to manage the symptoms and prevent complications of the disease. Acute episodes or illnesses may occur as part of a chronic disease. CF is a chronic disease. See also **acute**.

- **Cilia**

The tiny hair-like structures lining the cells of the airways. Cilia help move mucus and foreign matter, such as dust and bacteria, up and out of the lungs. Thick mucus, infection, tobacco smoke, and other irritants can damage the cilia.

- **Cirrhosis**

Fibrosis and scarring of the liver, which can be caused by a number of diseases. This is a complication that occurs rarely in CF. Cirrhosis in CF is caused by thick mucus blockage in the liver bile ducts. See also **bile duct**.

- **Clubbing**

A condition in which the ends of the fingers and toes are enlarged or bulblike. The nails become rounded. Clubbing can develop in people with CF. How severe the clubbing is does not always match how severe the lung disease is. It is not known why clubbing happens. Clubbing can sometimes be seen in other conditions including heart birth defects.

- **Colon**

Another name for the large intestine, a part of the digestive system. See also **intestines (small and large)**.

- **Colonization/Colonize**

The growth of bacteria and fungi in a part of the body where they will live over time. Everyone has bacteria that grow on the skin, in the mouth, and in the colon. However, it is not normal for a person to have bacteria grow in the airways. In CF, bacteria often live in the abnormal mucus in the airways.

- **Competent**

Having the ability, knowledge, and skills to carry out and complete tasks. Parents and their children with CF become competent CF managers after they have been trained and have been able to practice and successfully carry out CF management tasks. All children, with and without CF, work to become competent in developmental tasks or skills, such as getting dressed, playing a sport, or completing a homework assignment. See also **CF manager**; **developmental task**.

- **Complication(s)**

A problem caused by a disease. Common complications of CF are lung infections, malabsorption, and sterility.

- **Computed Tomography**

Also called CT. This is a type of radiology test that uses a computer to take pictures of an area of the body. The pictures show more detail than a simple X-ray.

- **Conceive(d)**

To become pregnant.

- **Conception**

When a man's sperm joins with a woman's egg to create a baby. The beginning of a pregnancy.

- **Congenital Bilateral Absence of the Vas Deferens (CBAVD)**

From birth, lacking the tubes that carry sperm from the testicles to the penis. This tube is called the vas deferens. If these tubes are missing or blocked, sperm cannot get into the semen and the male is infertile. See also **infertile**; **semen**; **testes**.

- **Constipation**

When a person does not have stools (bowel movements) regularly. Most people have at least one stool a day. A person who is constipated may not have a stool for several days. The stool may be harder than usual. Abnormal stool or reduced motion of the intestines can cause constipation. People can have constipation if they do not drink enough fluids or have enough fiber in their diets. Constipation is common in CF and some people with CF regularly need to use laxatives or stool softeners. See also **bowel movement (BM)**; **laxative**.

- **Contact Isolation**

Used in the clinic or hospital when a person gets certain types of infection that can be spread to other people. In CF, infections that require contact isolation include multiply resistant *Pseudomonas aeruginosa* and *Burkholderia cepacia*; methicillin-resistant *Staphylococcus aureus*; and certain viruses.

- **Contaminated**

When an item has something, such as germs, on or in it that can cause harm.

- **Contraception**

Methods used to prevent pregnancy.

- **Contraindicated**

A term applied to certain medicines or treatments when they are not to be used. For example, when a person has an allergy to certain antibiotics, those antibiotics are contraindicated.

- **Cor Pulmonale**

A complication that can be seen in people who have severe lung disease. Low oxygen levels and increased blood pressure in the lungs cause strain on the right side of the heart. The right side of the heart has to pump harder and can fail over time.

- **Corticosteroid(s)**

A type of steroid that is produced naturally in the body (in the adrenal gland) or is produced synthetically as a medication. Corticosteroid medicines are used to treat inflammation.

- **Cough**

The quick and forceful exhalation of a breath out of the lungs. Coughing is a natural body defense that helps clear the airways of foreign matter and mucus. People with CF need to have a strong cough to get mucus out of the lungs. Increased coughing is also a symptom of illness or a disease that is getting worse.

- **Crackle(s)**

An abnormal sound heard in the lungs. Usually due to damaged airways or excess mucus or fluid in the lungs.

- **Creatinine**

A waste product of the body that is usually gotten rid of by the kidneys in the urine. If the kidneys are not working properly, the blood creatinine level can go up. The blood creatinine level is a test to watch kidney function.

- **Cultures**

Lab tests done to grow germs that may be causing infections. Most often cultures are done to identify bacteria in a sample of body fluid, such as sputum (mucus). Bacteria may take several days to grow in culture. People with CF need to have regular sputum cultures to see what bacteria are present. Other cultures can be done to grow certain viruses or fungi. See also **sensitivity**.

- **Cyanosis**

A condition in which the skin or body looks blue. The blue color shows that the body doesn't have enough oxygen. A person may be described as "cyanotic." See also **hypoxia**.

- **Cystic Fibrosis**

Cystic fibrosis (or CF) is a chronic disease inherited from both parents. Abnormal (thick, sticky) mucus blocks the pancreatic ducts and the airways, and leads to the most common problems in CF: difficulty digesting food and lung infections. Treatments can improve symptoms and slow the progression of the disease.

D

- **Dehydrated/Dehydration**

A condition that results from a person not having enough fluid. Usually happens when a person is not drinking enough fluids and has extra fluid loss, such as with sweating or diarrhea. Dehydration can cause serious health problems. People with CF are at higher risk of dehydration.

- **Developmental Stage and Developmental Track**

Children develop physically, mentally, and socially in similar ways. A developmental stage is defined by physical and cognitive abilities, and emotional and social maturity common for a child of a certain age.

Developmental track refers to the developmental stages that a child progresses through in the order and at the same rate of other children. A child who is not on the developmental track would not be doing what other children of his or her age do.

- **Developmental Task**

A skill or behavior that a child learns by a certain age. Developmental tasks include speech; physical behaviors, such as walking or writing; and cognitive tasks, such as solving math problems or spelling.

- **DEXA Scan**

Short for dual-energy X-ray absorptiometry scan. An X-ray that checks bone thickness or density to diagnose bone disease. See also **osteoporosis**.

- **Diabetes**

A chronic disease caused by the pancreas not making enough insulin (insulin deficiency) and/or the body cells not responding normally to insulin (insulin resistance). Diabetes can be a complication in CF. Insulin is produced by the pancreas and controls how much sugar the body can use for energy and how much is stored. CF-related diabetes (CFRD) is usually treated with insulin. See also **CF-Related Diabetes (CFRD)**.

- **Diagnose(d)/Diagnosis**

Giving a name to a disease or illness that a person has. A health care provider makes a diagnosis based on a person's medical history, physical findings, and various test results.

- **Diaphragm**

The main breathing muscle—a dome-shaped muscle separating the chest and abdomen. People with CF may learn to use the diaphragm to cough better or make breathing easier.

- **Diarrhea**

Refers to watery stools. Diarrhea is most often caused by infection with certain bacteria or viruses. Other symptoms with diarrhea include fever, stomach pain, nausea, and vomiting.

- **Dietitian**

A health care professional who is trained to look at a person's diet and growth pattern. A dietitian can teach families how to adjust their children's diets so the children with CF get the calories and nutrients they need to be healthy. See also **nutrients**.

- **Differential Attention**

A way to modify or change behavior by ignoring unwanted behavior and paying attention to desired behavior. For example, a parent ignores her child when he whines (unwanted behavior), but as soon as the child talks without whining (desired behavior), the parent pays attention to him. If the parent is consistent about ignoring whining and paying attention to appropriate talk, the child will eventually stop whining.

- **Digest(ing)/Digestion**

The process of breaking down food into nutrients. The nutrients are absorbed into the bloodstream and used by the body. Digestion begins in the mouth and continues in the stomach and intestines.

- **Digestive System**

The mouth, esophagus, stomach, liver, gallbladder, pancreas, small intestine, large intestine (colon), rectum, and anus take in and digest food and remove the wastes not used by the body.

- **Digestive Tract**

See **gastrointestinal/GI tract**.

- **Distal Intestinal Obstruction Syndrome (DIOS)**

A partial or complete blockage of the intestine. The blockage is made up of abnormal stool with partly digested food and mucus. It is a severe form of constipation. DIOS is more common in CF as a person ages. Also called meconium ileus equivalent.

- **DNA**

Short for deoxyribonucleic acid. DNA strands are the chemical codes that make up genes. DNA determines the “genetic message” for each cell in the body. DNA is found in every cell.

- **DNase**

A type of protein that cuts long strands of DNA into small pieces. An inhaled drug used in CF called dornase alfa (Pulmozyme®) is a DNase. Dornase alfa breaks down DNA that gets in airway mucus from cells that have died. DNase helps make thick mucus thinner.

- **DNA Testing**

Since the pattern of DNA makes up the instruction code in each gene, genetic testing to look for CF gene mutations is sometimes called CF DNA testing. See also **DNA**; **genetic**.

- **Dosage**

The amount and the number of times per day medicine(s) should be taken as prescribed by a doctor.

- **Dose**

A measured amount of medicine given.

- **Drug Level**

A lab test to measure the amount of a drug in the blood. Drug levels help make sure the dosage is high enough to work, but not too high that it causes side effects. Drug levels are needed for certain medicines used in CF, such as aminoglycoside antibiotics. See also **aminoglycoside antibiotic**; **peak level**; **trough level**.

- **Dry Powder Inhaler**

DPI for short. This device delivers a very fine powder of medicine that a person breathes in.

- **Ducts**

Passages or tubes in the body. Ducts from the pancreas carry enzymes to the small intestine to help digest food. In CF, the ducts are often blocked with abnormal mucus. The person with CF then needs to take replacement enzymes.

- **Dyspnea**

Shortness of breath. A feeling that a person cannot get enough air.

E

- **Edema**

A buildup of fluid in tissues that causes swelling or congestion. Edema may show up as swollen feet or ankles. Pulmonary edema occurs in the lungs and can be seen with cor pulmonale or other causes. See also **cor pulmonale**.

- **Ejaculated/Ejaculation**

The act of sperm and semen leaving the body through the penis. See also **penis**; **semen**; **sperm**.

- **Elastase**

One of the enzymes produced in the pancreas. Elastase helps break down some proteins in food in the intestine. It can also be measured in the stool to diagnose pancreatic insufficiency. See also **enzymes**; **pancreatic enzymes**; **pancreatic insufficiency**; **protein**.

- **Electrolytes**

Chemical salts that help cells work. Electrolytes include sodium, chloride, potassium, and bicarbonate. See also **bicarbonate**; **chloride**; **potassium**; **sodium**.

- **Embolization**
A procedure to stop bleeding in the lungs. A long thin tube (catheter) is threaded to the blood vessels around the airways. Little plugs of plastic-like material are injected to block a leaky blood vessel. This is done sometimes in CF when a person has severe hemoptysis. See also **hemoptysis**.
 - **Endocrinologist**
A doctor with special training in the diagnosis and treatment of diabetes and other diseases, including problems with thyroid hormone and growth hormone. See also **CF-related diabetes; diagnosis; growth hormone**.
 - **Endoscopy**
A test that uses a tube with a fiber-optic camera and suction to look inside the body. For example, an endoscope can be used to look into the esophagus and stomach or the colon (large intestine). The doctor can get biopsies (small samples of tissue) and fluid samples.
 - **Endurance**
The ability or strength to continue or last. Can be referred to as a person's "staying power." Sticking to or persevering with a task.
 - **Enema**
Putting fluid up through the anus into the rectum and lower colon. See also **anus; colon; rectum**.
 - **Enzymes**
Chemicals the body uses to break down and digest food. These enzymes are called digestive or pancreatic enzymes. Enzymes produced by the pancreas include lipase, protease, and amylase. Without enzymes, food cannot be broken down into nutrients the body can use. In CF, mucus may block the ducts or tubes that carry enzymes. Most people with CF must take pancreatic enzymes (also called replacement enzymes or enzyme supplements) to digest food. See also **malabsorption; pancreatic enzymes**.
 - **Enzyme Supplements**
See **pancreatic enzymes**.
 - **Esophageal Varices**
Blood vessels in the esophagus that get enlarged and swollen because of pressure and blood backup with cirrhosis. These blood vessels can break open and bleed, which may cause a person to vomit up blood. See also **cirrhosis; esophagus; liver**.
 - **Esophagitis**
An inflammation or irritation of the esophagus, most often caused by gastroesophageal reflux (GER). Esophagitis can cause heartburn (pain in the chest). See also **gastroesophageal reflux**.
 - **Esophagus**
The tube that connects the throat to the stomach.
 - **Estrogen**
A hormone that comes from the ovaries in females. Estrogen is important in female sexual development and function. See also **ovaries**.
 - **Exacerbation**
A time when a person is having more signs and symptoms of illness or disease problems that need treatment. In CF, people can have pulmonary exacerbations. See also **pulmonary**.
 - **Exhale(ing)/Exhalation**
Breathing out; the flow of air out of the lungs with each breath.
 - **Exocrine Glands**
Tissues in the body that make secretions. The sweat, salivary, and tear glands are exocrine glands. In the airways and pancreas, exocrine glands produce mucus. In CF, these glands make abnormal secretions. See also **sweat glands**.
 - **Expiration/Expiratory**
The time when a person is exhaling (air flowing out of the lungs) during breathing. See also **exhale(ing)/exhalation; FEV1**.
- F


- **F508del**
Also written as deltaF508 or ΔF508. The most common gene mutation (abnormal gene) that has been found to cause CF. See also **gene(s)/gene mutation; mutation(s)**.
 - **Face Mask**
Babies and young children need to wear a face mask to inhale medications through the nose and mouth as a mist during breathing treatments. The mask is connected to a nebulizer cup and air compressor machine. The mask makes it possible for the baby or young child to get more of the medication into his or her lungs.

- **Failure to Thrive**
A child who is not growing normally. Failure to thrive is a common problem seen in CF.
- **Fallopian Tubes**
The tubes that connect the ovaries to the uterus. The woman's eggs travel from the ovaries through these tubes to the uterus. See also **ovaries**; **uterus**.
- **Fast/Fasting**
Nothing to eat or drink for a number of hours.
- **Fat**
A type of nutrient found in many foods and an important source of energy for the body. Most fats have to be digested by pancreatic enzymes in the intestine.
- **Fat-Soluble**
Able to be dissolved in a fat or oil. Fat-soluble vitamins (A, D, E, and K) are not absorbed well in CF so extra vitamin supplements are needed.
- **Fecal**
Being of or related to stool (feces). See also **feces**.
- **Fecal Elastase**
Elastase is an enzyme that is normally found in stool. The elastase level in the stool is low in a person with CF who has pancreatic insufficiency. It can be measured to test for malabsorption. See also **pancreatic insufficiency**; **malabsorption**.
- **Feces**
Another word for stools or bowel movements. See also **bowel movement (BM)**.
- **Fellow**
A doctor who has completed a residency and is doing additional training to become a specialist in an area such as pediatric lung diseases (pediatric pulmonologist). Fellows care for patients under the supervision of a specialist physician.
- **Fertile**
Able to get pregnant (woman) or cause pregnancy (male). See also **infertile**.
- **Fertility**
The ability to conceive a baby.
- **Fertilize**
When a man's sperm joins with a woman's egg.
- **FEV₁**
Short for forced expiratory volume in 1 second. During spirometry, this measures the amount of air that a person blows out in the first second of exhaling forcefully. If a person's FEV₁ is average for sex, age, height, and race, it is said to be 100 percent predicted. See also **exhaling**; **expiration/expiratory**; **percent predicted**; **spirometry**.
- **Fibrosis**
Formation of fiber or scar tissue. In CF, fibrosis can occur in the pancreas, lungs, or liver.
- **Flatus**
Gas produced in the intestine that is passed from the anus. Commonly called a fart or toot.
- **Flutter[®] Device**
A handheld device that is used during airway clearance to help move mucus out of the lungs. A person with CF blows into the device, which loosens mucus by making vibrations in the airways. He or she needs to cough between blows. A CF health care team member teaches people with CF and families how to use the Flutter[®] device.
- **Fundoplication**
A surgical procedure used to tighten the muscle at the upper end of the stomach where it joins the bottom of the esophagus. The procedure is used to control gastroesophageal reflux (GER) when medical treatments have failed. See also **gastroesophageal reflux**.
- **Fungus**
A group of plants that have no leaves or flowers. Mildews, molds, and mushrooms are forms of fungi. Some people develop allergy sensitivity to *Aspergillus* mold. Rarely, certain fungi can cause infection in the lungs. *Candida* is another common fungus. *Candida* can cause thrush in the mouth or a diaper rash. See also **Aspergillus**; **Candida**; **colonization/colonize**.
- **FVC**
Short for forced vital capacity. Largest amount of air that a person can breathe in and out with a good effort. One of the measures done by a spirometry test. See also **spirometry**.

G

- **Gallbladder**

The gallbladder is a small organ attached to the liver and is part of the gastrointestinal tract. The gallbladder stores extra bile the liver produces. Bile fluid is released from the liver and helps digest food in the intestines. See also **bile**; **gastrointestinal/GI tract**.

- **Gallstone**

A stone-like mass that can be found in the gallbladder or bile duct. See also **bile duct**; **gallbladder**.

- **Gas Exchange**

The exchange of carbon dioxide for oxygen. Gas exchange takes place in the alveoli or air sacs at the end of the smallest branches of the airways (bronchioles). Oxygen passes through the walls of the alveoli into the bloodstream as carbon dioxide moves from the bloodstream into the alveoli. From the alveoli, the carbon dioxide passes into the bronchi and moves out of the lungs as a person breathes out, or exhales.

- **Gastric Fluid**

An acidic fluid produced in the stomach that helps digest food. As the stomach churns, the gastric fluid mixes with the food and helps break it down.

- **Gastroenterologist**

A doctor who has special training in the diagnosis and treatment of diseases of the digestive system, including the esophagus, stomach, intestines, and liver. See also **digestive system**; **esophagus**; **intestines**; **liver**.

- **Gastroesophageal Reflux**

Also called GE reflux or GER for short. The flow of stomach contents back up into the esophagus. Gastroesophageal reflux can cause vomiting, heartburn, and esophagitis. Tests used to diagnose GER include X-rays, an endoscopy, or a 24-hour pH probe. Medicines can be used to try to control GER. Sometimes a surgery is required called a fundoplication. See also **endoscopy**; **esophagitis**; **fundoplication**; **pH probe**.

- **Gastrointestinal**

GI for short. Relating to the stomach and intestines. Parents of children with CF keep track of gastrointestinal or digestive symptoms (such as the size and appearance of stools), to monitor how well their child's enzymes and high-calorie diet are working. See also **digestive system**; **gastrointestinal/GI tract**; **malabsorption**.

- **Gastrointestinal/GI Tract**

Gastro = stomach; intestinal = intestines. The gastrointestinal or GI tract (sometimes also called the digestive tract) refers to the entire digestive system—the pathway food travels through the body. In the GI tract, the food is digested and the nutrients absorbed by the body. The waste from food passes out of the body through the rectum and anus, which are located at the end of the GI tract. See also **digestive system**.

- **Gastrostomy Tube or Button**

GT or G-button for short. A tube that is placed through the abdominal wall into the stomach to provide nutrition. See also **abdominal wall**.

- **Gene(s)/Gene Mutation**

The basic unit of heredity. Genes determine each person's physical and chemical makeup. Genes are carried on chromosomes. A change in a gene is called a gene mutation. CF is caused by an abnormal gene mutation on the chromosome number 7. See also **autosomal recessive**; **chromosomes**; **genetic**; **mutation(s)**.

- **General Anesthesia**

When a person is given medicines that put him or her into a sleep-like state for surgery, tests, or procedures. This is usually done by a specialist physician called an anesthesiologist. A person who is under general anesthesia is not aware of what is happening, does not feel pain, and may need support to breathe. The person often does not remember anything that happened during the time he or she was under anesthesia.

- **Generic**

A substitution for a specific brand of medication. Generic medications do not have a registered trademark name. Some generic medicines work as well as the brand name versions. However, for CF care, it is recommended that families avoid generic enzymes because they do not work as well.

- **Gene Therapy**

Research on future treatments for CF includes gene therapy in which healthy genes take the place of abnormal genes. The research focuses on how to get the healthy genes to the cells to take over for the abnormal genes. The first step in gene therapy was the discovery of the CF gene in 1989.

- **Genetic(s)**

Anything related to genes and inheritance (heredity). CF is a genetic disease. See also **autosomal recessive**; **chromosomes**.

- **Genetic Counselor/Counseling**

A medical professional trained in genetics. A discussion (or counseling) with a genetic counselor can help a person understand the risk of passing on an inherited disease to his or her children. For example, families with a history of CF may learn their risk of having a child with CF.

- **Genetic Mutation**

See **gene(s)/gene mutation**.

- **Genitals**

The male or female body parts involved in reproduction (the sexual organs).

- **Germs**

Living organisms (microbes) that often cause infection or disease. Examples include bacteria and viruses. See also **microorganisms; organism(s)**.

- **Glucose**

A type of sugar that is found in many foods. An important nutrient used by all cells for energy. Blood glucose levels are high if a person has diabetes. See also **blood glucose**.

- **Growth Charts**

Graphs that show normal patterns of growth for boys and girls at various ages. A person's own measures, such as height and weight, can be put on the graph to compare to others and to track over time.

- **Growth Hormone**

A hormone that is made in the brain. It is important in normal growth, including growth of bones and muscles.

- **Gut Flora**

Helper bacteria that normally live in the body's intestines. These bacteria help break down undigested carbohydrates that are passing through the intestines. They also help the intestines stay healthy and develop. They block the growth of bacteria that can cause infections, such as diarrhea. Other "helping tasks" include producing vitamins (such as vitamin K), helping the immune system, and preventing inflammation in the intestines.

- **Gynecologist**

A doctor who has special training in diagnosing and treating the female reproductive system, including the uterus, cervix, ovaries, and fallopian tubes. See also **cervix; fallopian tubes; ovaries; uterus**.

H

- **Haemophilus Influenzae**

Also called *H. influenzae*, for short. A type of bacteria that may cause ear infections, pneumonia, and meningitis in young children. *H. influenzae* can colonize the airways of a person with CF and can cause bronchitis. One type of *Haemophilus influenzae*, type b (often called Hib), can now be prevented with a vaccine. The "flu" is caused by the influenza virus, not by *H. influenzae* bacteria. See also **bacteria; bronchitis; colonization/colonize**.

- **Hand Hygiene**

Cleaning hands to prevent spread of germs.

- **Heat Stroke/Heat Exhaustion**

Serious illness caused by overexposure to heat—often seen together with dehydration. Heat exhaustion is less severe than heat stroke. Without medical treatment and immediate cooling down, a person can die from heat stroke. People with CF may be at higher risk of heat-related problems.

- **Hemoptysis**

Coughing up blood from the lungs. Bleeding can occur in a person with CF who has lung disease and infection. Bleeding often stops on its own. A procedure called an embolization may be needed to plug the bleeding blood vessel. See also **embolization**.

- **Hereditary**

A trait or condition that is passed genetically (inherited) from parents to children. See also **genetic**.

- **Heterozygote**

A person who has inherited two different genes for a trait or disease. A person who is a heterozygote for CF has two different CF gene mutations. See also **carrier**.

- **High-Calorie Food Additives**

Products added to food to increase the number of calories. Children with CF need more calories than other children their age to absorb enough nutrients to grow and be healthy. Examples of high-calorie food additives include polycose powder, MCT oil, and corn oil.

- **High-Calorie Supplements**

Liquids or puddings that are high in calories. High-calorie supplements are given by mouth or through a gastrostomy tube to help a person gain or maintain weight. See also **gastrostomy tube**.

- **High Frequency Chest Wall Oscillation**

Also called percussive vest therapy or HFCWO for short. A type of airway clearance used to help loosen mucus in the lungs. A person wears an inflatable vest attached to a machine that causes the vest to vibrate rapidly. The machine has various pressure settings. A CF health care team member teaches people with CF and families how to use HFCWO. See also **percussive vest**.

- **Holding Chamber**

Also called a spacer. A device that is used with a metered dose inhaler (MDI) to hold the puff of medicine so that a child can inhale it more slowly and deeply into his or her airways.

- **Homozygote**

A person who has inherited two of the same genes for a particular trait or disease. A person who is a homozygote for CF has two of the same CF gene mutations.

- **Hormone**

A substance that is produced by endocrine cells in the body and affects body functions. Examples include insulin and growth hormone. See also **growth hormone; insulin**.

- **Huff(ing)**

A way to move mucus out of the lungs. Huffing is a modified type of cough. It is similar to the way people breathe when they fog up a mirror with their breath. A person with CF breathes out actively, blowing the air out with force. A member of the CF health care team will teach him or her how to “huff.”

- **Hyperglycemia**

High blood sugar. A person with CF can have high blood sugar because of CF-related diabetes or certain medicines. See also **CF-related diabetes (CFRD); insulin**.

- **Hyperinflation**

When one or both lungs are overfilled with air. Air can get trapped behind mucus in the lungs of a person with CF. This may be seen on a chest X-ray.

- **Hypertonic Saline**

A solution that is high in sodium chloride (salt) content. Hypertonic saline (Hyper-Sal™) is used in CF to help clear mucus from the airways. Usually a 7% solution is inhaled. Some people cannot tolerate hypertonic saline due to bronchospasm. See also **bronchospasm**.

- **Hypoxia**

Too little oxygen in the body. People with CF can have hypoxia when they have lung problems.



- **IgE**

Short for immunoglobulin E. A protein that is involved in allergic reactions. People with allergies have a higher than normal blood IgE level. The level of IgE is used to check for ABPA. See also **ABPA**.

- **Ileum**

The last part of the small intestine that connects to the large intestine. See also **intestines**.

- **Immune System**

The system in the body that prevents disease and fights infections. The immune system contains several parts including white blood cells, antibodies, and organs, such as the spleen. See also **antibodies**.

- **Immunizations**

Also called vaccinations. Used to help the body develop antibodies against certain bacterial or viral infections. A number of immunizations are recommended for all children and adults. People with CF should get a yearly influenza vaccine. See also **antibodies**.

- **Immunoreactive Trypsinogen (IRT)**

This is a test that is used in newborn screening for CF. Most infants with CF will have a high IRT level.

- **Impaired Glucose Tolerance**

When blood glucose levels are higher than normal, but not high enough to diagnose diabetes. See also **blood glucose; diabetes; diagnose**.

- **Infection(s)**

What happens when germs grow in the body, often causing harm. Infections can be acute or chronic. An infection can vary in how severe it is from person to person. A person can have more than one infection at the same time. Infections can cause a number of symptoms, such as fever, cough, or diarrhea.

- **Infertile**

Unable to get pregnant (female), or to cause pregnancy (male). See also **infertility**.

- **Infertility**

When a person is not able or finds it very difficult to conceive or produce children. More than 90 percent of men with CF are infertile because the vas deferens (the tube that carries sperm) is blocked. Women with CF can conceive, but may find it harder to get pregnant because of abnormal mucus blocking the cervix. See also **vas deferens**.

- **Inflammation**

Swelling of the body's cells because of an infection or injury. Inflammation can be a normal reaction that helps clear the problem and repair injured cells. Too much inflammation, however, can cause problems. In CF, chronic inflammation in the lungs causes damage over time.

- **Influenza**

Also called “the flu” for short. An acute infection caused by an influenza virus. There are two types of influenza viruses that cause the infection: types A and B. Common flu symptoms include headache, muscle aches, fever, chills, sore throat, and cough. A yearly influenza immunization (flu shot) helps protect people from getting the flu. See also **immunizations**.

- **Inhalations**

The number of puffs or doses of medicine to be inhaled or breathed in.

- **Inhale(ing)/Inhalation/Inspiration**

Breathing in; the flow of air into the lungs.

- **Inhaler**

Another name for metered dose inhaler or puffer. See **metered dose inhaler**.

- **Inherit(ed)**

A trait or disease that is passed from parents to children. See also **hereditary**.

- **Inpatient**

A person admitted to the hospital for treatment.

- **Insulin**

A hormone produced by cells in the pancreas that helps the body use certain sugars including glucose. A person who does not have enough insulin has diabetes. Some people with diabetes can make insulin, but the cells do not respond to it normally. Some people with CF develop CF-related diabetes (CFRD). See also **CF-related diabetes (CFRD); hormone**.

- **Intestines (Small and Large)**

Also called the bowels. The hollow tubes that are found in the abdomen. Food passes from the stomach into the small intestine where most of the digestion in the body takes place. Enzymes from the pancreas pass through ducts into the small intestine to help digest or break down food so that it can be absorbed by the body. Any undigested food then travels to the large intestine (also called the colon) where good bacteria help turn the leftover waste from the food into stool (bowel movement). The large intestine helps move the waste or stool out of the body through the rectum and anus. See also **digestive system; pancreas**.

- **Intrapulmonary Percussive Ventilation (IPV®)**

IPV® is another way to do airway clearance. Mucus is loosened by making vibrations in the airway. The IPV® machine delivers very fast short pulses (bursts) of air through a mouthpiece. See also **airway clearance**.

- **Intravenous(ly)**

IV for short. Given in a vein. A small tube (also called a catheter) is placed into a vein to give medicines.

- **Intussusception**

A problem in which a part of the intestine telescopes into another part causing a blockage. Usually this occurs right where the small intestine joins the large intestine.

- **Iron**

A mineral needed by the body. It is used by red blood cells. Low iron levels can cause anemia.

- **Irritants**

Substances that can cause inflammation when they come in contact with the body. Irritants that a person breathes in, such as air pollutants or tobacco smoke, can trigger inflammation and bronchospasm. See also **bronchospasm; inflammation**.

- **IV**

Short for intravenous. Usually it refers to an IV catheter, a small tube placed in a vein. See also **central line; intravenous(ly); peripheral line; PICC line**.

K

■ Kidney Function Tests

Blood tests that help show whether the kidneys are working well. Kidney irritation or injury from infection or certain medicines will cause abnormal blood levels of blood urea nitrogen (BUN) and creatinine. These are also called renal function tests. Kidney function tests are monitored when a person with CF is treated with certain medicines such as aminoglycoside antibiotics. A urine sample is also looked at to see if there is irritation of the kidneys. See also **creatinine**.

L

■ Lactose Intolerance

Problems digesting lactose, a type of sugar found in certain foods, such as cow's milk and other dairy products. Lactose intolerance is a common problem among all children. Signs of lactose intolerance include diarrhea, gas, and/or bloating right after eating or drinking products that contain cow's milk.

■ Large Intestine

Also called the colon. See **intestines (small and large)**.

■ Laxative(s)

Medicines that help treat or prevent constipation and move stool through the intestines. See also **constipation; intestines**.

■ Lipase

One of the enzymes produced in the pancreas. Lipase helps break down fats in food in the intestine. Lipase is included in pancreatic enzyme supplements in CF. See also **enzymes; pancreatic enzymes**.

■ Liver

An organ of the body located in the upper right side of the abdomen just below the diaphragm. The liver does many things. It is involved in the breakdown of protein and carbohydrates in the blood. The liver also produces bile fluid that goes to the small intestine and helps with digestion.

■ Liver Enzymes

Chemicals that work in the liver to help break down nutrients and medicines from the blood. Blood levels of liver enzymes show how well the liver is working and if it is injured.

■ Liver Function Test

A group of blood tests that help tell if the liver is working normally. Results may be high if the liver is not working well, or has an infection. For people with CF, liver function tests are done at least once a year. See also **liver**.

■ Lobes

The lungs are divided into sections called lobes. The right lung has three lobes: upper, middle, and lower. The left lung is slightly smaller because the heart sits in the left side of the chest. The left lung has two lobes: upper and lower. When families learn how to do postural drainage and percussion (PD & P), they learn about the lobes of the lungs so that they know where to clap their child with CF to help move mucus out of the lungs.

■ Lower Respiratory Tract

The windpipe (trachea), lungs, and airways below the vocal cords.

■ Lower Respiratory Tract Infections

Infections, such as bronchitis or pneumonia, involving the airways and/or lung tissue. In CF, the most common bacteria that cause these infections are *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and *Haemophilus influenzae*. Viruses that may cause the infections include respiratory syncytial virus (RSV), adenovirus, and influenza (the flu). Bacterial infections are treated with antibiotics, but there are no medicines for most viral infections.

■ Lung Function

How the lungs work. Lung function includes how well air moves in and out, and how well the lungs bring in oxygen and blow out carbon dioxide. Lung function can be measured by pulmonary function tests. See also **pulmonary function tests**.

■ Lung Function Tests

See **pulmonary function tests**.

■ Lung Infection(s)

Lung infections in CF are most often caused by bacteria and/or viruses. Bacteria grow well in the thick abnormal mucus in the airways and sinuses in CF. Infections caused by bacteria need to be treated with antibiotics. See also **bacteria; lower respiratory tract infections; viruses**.

- **Lungs**

The lungs provide oxygen for the whole body. The trachea branches into the bronchi or major airways in the right and left lungs. The bronchi then branch off into the smallest airways called the bronchioles. The bronchioles end deep in the lungs with clusters of alveoli or air sacs where gas exchange takes place. See also **airway(s)**; **alveoli**; **bronchi**; **bronchioles**; **gas exchange**; **trachea**.

- **Lung Tissue**

Lung tissue includes the alveoli (air sacs) and parts of the lungs around the air sacs. Infections, such as pneumonia, can occur in the lung tissue. See also **alveoli**.

M

- **Magnesium**

A mineral in the diet that is needed by the body. Low levels can occur at times in CF.

- **Malabsorption**

A condition in which the body does not absorb nutrients normally from the intestines. Most people with CF have malabsorption from pancreatic insufficiency. Without pancreatic enzymes, they do not digest food properly. See **pancreatic insufficiency**.

- **Malnutrition**

A condition in which the body is not getting good nutrition and a person's weight is too low. A child is not growing as expected. Can be caused by malabsorption, not taking in enough calories and nutrients, and increased demand from illness. Malnutrition is a serious complication in CF.

- **Manual Percussor**

A plastic cup-like device that a person holds in his or her hand to do airway clearance (percussion) on the chest.

- **Mechanical Percussor**

An electric device that is held in the hand to deliver thumps or vibrations to the back and chest for airway clearance. See also **airway clearance**.

- **Meconium**

A thick, dark substance normally found in a baby's intestine. It is the first stool passed after birth.

- **Meconium Ileus**

A condition seen in 10 to 15 percent of newborn babies with CF. The intestine is blocked with a mixture of meconium and mucus. An operation may be needed to treat the blockage. All infants with CF who have meconium ileus have pancreatic insufficiency. See also **meconium**; **pancreatic insufficiency**.

- **Meconium Ileus Equivalent**

See **distal intestinal obstruction syndrome (DIOS)**.

- **Medicine Syringe**

A plastic tube with a plunger and markings to measure liquid medicine. The medicine syringe does not have a needle. The plunger allows parents to squirt medicine inside their child's cheek so that he or she does not choke on the medicine. A baby can suck on the medicine syringe like a nipple.

- **Menses**

The menstrual period blood flow. Also called a period. See also **menstrual cycle**; **menstrual periods**.

- **Menstrual Cycle**

The monthly series of changes that occur in women. Starts with the release of an egg from the ovary and, if no conception occurs, ends with the menses. See also **conception**; **menses**; **ovaries**.

- **Menstrual Periods**

The time in the menstrual cycle that the blood flows out of the uterus. Also called a period. Can be irregular in females who are underweight. See also **menses**; **menstrual cycle**; **uterus**.

- **Menstruate**

See **menses**; **menstrual periods**.

- **Metered Dose Inhaler**

MDI for short. A device that delivers a dose of medicine as a puff for a person to inhale. A holding chamber (spacer) can be used with an MDI to help a person with CF breathe the medicine in more slowly and deeply. An MDI is also referred to as an inhaler or puffer. See also **holding chamber**.

- **Methicillin-Resistant Staphylococcus Aureus**

MRSA for short. A type of bacteria that can cause infection. MRSA is a strain of *Staphylococcus aureus* that is more resistant to antibiotics. MRSA has caused skin and bone infections and is found at times in the airways of people with CF. See also **bacteria**.

- **Microorganisms**

Very small life forms, such as viruses, bacteria, and fungi, that can only be seen through a microscope (cannot be seen with the naked eye). Some microorganisms can cause disease or infection. See also **bacteria**; **fungus**; **viruses**.

- **Milk Drip(s)**

Also called tube feedings. A way to get high-calorie supplements using a gastrostomy or nasogastric tube. A milk drip may be done at night or while a person is asleep. See also **gastrostomy tube**; **nasogastric tube**.

- **Minerals**

Substances found in the diet that are important to the body. There are a number of minerals that the body uses. These include the salts (sodium and chloride), calcium, magnesium, and zinc. A person with CF may need extra amounts of certain minerals.

- **Mouthpiece**

A child 5 years and older can learn to use a mouthpiece instead of a face mask to inhale medicine as a mist during breathing treatments. The mouthpiece is used with a nebulizer and air compressor machine.

- **Mucolytic Medicines**

Medicines used to break down mucus. Used at times in CF to help with thick mucus in the airways or in the intestines. See also **DNase**.

- **Mucous Membranes**

The tissues in the body that produce mucus. The mucous membranes line various passages of the body. For example, they line the nose, mouth, sinuses, airways, stomach, and intestines.

- **Mucus**

A liquid produced by the glands in the body. Usually it is thin and slippery. Mucus moistens and protects the airways and GI tract. In people with CF, the mucus is thick and sticky. It can block the airways, sinuses, and intestines. When mucus is spit up or coughed out it is called sputum. See also **sputum**; **sputum culture**.

- **Mutation(s)**

A permanent change in genetic material that usually involves a single gene. There are more than 1000 gene mutations on the 7th chromosome that have been found in people with CF. The most common gene mutation found in CF is called F508del (deltaF508 or Δ F508). See also **chromosomes**; **F508del**; **gene(s)/gene mutation**.

- **Mycobacteria**

See **AFB**.

N

- **Nasal Polyp**

A tissue growth that is found in the nasal passage. Usually develops because of inflammation from infection or allergy. One or more polyps can occur in CF and may need to be removed by surgery. New polyps can develop even after surgery.

- **Nasogastric Tube**

NG tube for short. A tube that is placed into the nose and down the esophagus to the stomach. The tube is used to remove air or stomach fluid, or to deliver nutrition. See also **esophagus**.

- **Nebulizer**

A plastic device that holds liquid medicine for breathing treatments. When an air compressor blows air into the nebulizer, the liquid turns into a mist. A person with CF inhales the mist using a mask or mouthpiece. See also **aerosol**.

- **Nurse Practitioner**

A nurse who has an advanced degree with more education than other types of nurses. A nurse practitioner is trained and licensed to practice under the supervision of a physician. This includes conducting physical examinations and prescribing medications.

- **Nutrients**

Substances the body uses from digested food for energy, growing, and normal functioning. Nutrients fall into five groups: proteins, carbohydrates, fats, vitamins, and minerals.

O

- **Obstetrician**
A doctor who cares for women who are pregnant through the birth of the baby.
- **Obstruction**
A blockage of a tube or passageway, such as the airways or intestines. In CF, the airways become blocked with abnormal mucus. Obstruction can also be caused by bronchospasm of the airways. Abnormal stool can block the intestines. See also **bronchospasm**; **distal intestinal obstruction syndrome (DIOS)**.
- **OMNI Scale of Perceived Exertion**
A measure that shows increasing degrees of effort that a person feels during exercise. Can be used to rate a person's level of work (exertion) during physical activity.
- **Oral/Orally**
By mouth.
- **Oral Glucose Tolerance Test (OGTT)**
A test to diagnose diabetes or impaired glucose tolerance. It is usually done after a person has had nothing to eat or drink for 12 hours. Blood samples are taken before and up to 2 hours after drinking a set amount of glucose. See also **diabetes**; **glucose**; **impaired glucose tolerance**.
- **Oral Temperature**
To take a person's temperature by placing a thermometer in his or her mouth. Children aged 4 years and older usually can have their temperatures taken orally.
- **Organism(s)**
A living thing. Often used to refer to bacteria or germs.
- **Oropharyngeal Flora**
Bacteria that grow normally in the mouth and throat. See also **bacteria**.
- **Oscillating Positive Expiratory Pressure**
A type of positive expiratory pressure (PEP) device used in airway clearance. In addition to the expiratory pressure, the device makes vibrations (oscillations) in the airways to loosen mucus. See also **airway clearance**; **expiration/expiratory**; **PEP device**; **positive expiratory pressure**.
- **Osteoporosis**
When bones are less thick or dense and are weak. People with osteoporosis have a higher risk for bone breaks (fractures). See also **bone density**; **DEXA scan**.
- **Otolaryngologist**
A doctor who has special training in diseases of the ears, nose, throat, and upper airway. Also called an ENT (ear, nose, and throat) specialist.
- **Ototoxicity**
Irritation or damage to the inner ear that affects hearing or can cause symptoms, such as ringing in the ears. Can be a side effect of certain medicines (for example, aminoglycoside antibiotics). See also **aminoglycoside antibiotic**.
- **Outpatient**
A person being treated in a clinic, doctor's office, or emergency center and not admitted to the hospital.
- **Ovaries**
Part of the female reproductive system where eggs and hormones (including estrogen) are made. There are two ovaries connected by the fallopian tubes to each side of the uterus. See also **estrogen**; **fallopian tubes**; **ovulation**; **uterus**.
- **Ovulation**
The process of an ovary releasing an egg into the fallopian tube. This occurs during the menstrual cycle. See also **fallopian tubes**; **menstrual cycle**; **ovaries**.
- **Oximetry/Pulse Oximetry**
A test that measures oxygen saturation in the blood. A device is applied to the skin that uses a beam of light to measure oxygen and does not hurt. See also **pulse oximeter**.
- **Oxygen Saturation**
The result of an oximetry test, given as a percentage. Tells how much of the hemoglobin in the red blood cells is carrying oxygen. When all of the hemoglobin is full, it is "saturated." If the test result is low, a person may need more oxygen.

P

■ PaCO₂

A measure of the amount of carbon dioxide in the blood. PaCO₂ stands for the partial pressure of carbon dioxide (CO₂). Carbon dioxide is a waste gas produced by the cells that the body must get rid of. Normally, a person gets rid of carbon dioxide by breathing it out of the lungs. The PaCO₂ level tells how well the lungs are able to remove carbon dioxide. A high PaCO₂ is a sign of severe respiratory problems.

■ Pancreas

A digestive gland located behind the stomach and connected to the small intestine. The pancreas produces enzymes that pass into the small intestine to digest food. In CF, the ducts (tubes) in the pancreas may be blocked by mucus and keep enzymes from getting to the small intestine. A lack of pancreatic enzymes causes malabsorption. The pancreas also produces insulin that passes into the bloodstream to control glucose (sugar). See also **malabsorption**.

■ Pancreatic Enzymes

People with CF need to take pancreatic enzymes (also called replacement enzymes or enzyme supplements) because the enzymes made by the pancreas cannot get through the blocked ducts (tubes) to the small intestine to help digest food. Pancreatic enzymes are taken by mouth (in the form of beads, powder, or capsules) to help break down fats and proteins that are important for growth and good nutrition. See also **enzymes**; **malabsorption**.

■ Pancreatic Insufficiency

A condition in which a person does not have enough enzymes being delivered from the pancreas to the intestines for digestion. Most people with CF have pancreatic insufficiency. This causes malabsorption. See also **digest(ing)/digestion**; **enzymes**; **malabsorption**; **pancreatic enzymes**.

■ PaO₂

A measure of the amount of oxygen in the blood. PaO₂ stands for the partial pressure of oxygen (O₂). A low PaO₂ means that not enough oxygen is getting from the lungs into the blood.

■ Particle

A very small drop of something. Inhaled medicines are made of small particles that form a mist from a nebulizer or from an inhaler.

■ Pathogens

Germs that cause disease or infection. Can include viruses, bacteria, and fungi. See also **bacteria**; **fungus**; **viruses**.

■ PD & P

See **postural drainage and percussion**.

■ Peak Level

The highest level of a medicine in the bloodstream as measured by a drug level test. Some medicines must reach certain peak levels to be effective. Peak levels are measured to make sure the right dose is being given to work safely and well.

■ Pediatrician

A physician (doctor) who has completed additional training after medical school in the medical care of children and in working with families.

■ Pediatric Nurse Practitioner

A nurse practitioner who cares for children. See also **nurse practitioner**.

■ Pelvic Exam

An exam that is done through the vagina to assess the female reproductive organs, including the vagina, uterus, and ovaries. See also **ovaries**; **uterus**; **vagina**.

■ Penis

The male organ through which urine and semen leave the body.

■ PEP Device

Short for positive expiratory pressure device. A small device used for airway clearance to loosen mucus. Breathing out into the PEP device causes vibrations in the airways. A CF health care team member teaches people with CF and families how to use a PEP device. See also **positive expiratory pressure**.

■ Percentile

It is a value at or below which a certain percentage of the whole group falls. So on a growth chart for height — the 20th percentile means that 20 percent of people are at the same height or shorter, while the 99th percentile means that only 1 percent are taller and 99 percent are the same height or shorter. See also **growth charts**.

- **Percent Predicted**

A value that is based on people in a group. For example, lung function test results are often compared to those of people of the same sex, age, height, and race. If a person's FEV₁ is average, it is said to be 100 percent predicted. See also **FEV₁**; **lung function**.

- **Percussion**

A technique used with postural drainage for airway clearance. A cupped hand or plastic cup-like device is clapped over the rib cage. This sends a force through the lungs to loosen mucus. Percussion of different areas of the chest and back helps move mucus up into the airways so that it can be coughed out. A CF health care team member teaches families how to do percussion and postural drainage. See also **postural drainage and percussion**.

- **Percussive Vest**

Another way to do airway clearance. By wearing a vest attached to a machine, the child coughs out mucus that has been loosened by the vest shaking or vibrating the chest. The CF health care team teaches families how to use the percussive vest. See also **high frequency chest wall oscillation**.

- **Perforation**

A sudden hole or tear in a part of the body.

- **Peripheral Line**

An IV (intravenous) line in a small vein, usually in the arm or hand. A short catheter (tube) is inserted into the vein to deliver fluid and medicines. A peripheral IV is farther from the heart than a central line. See also **central line**.

- **PFTs**

Short for pulmonary function tests. See **pulmonary function tests**.

- **pH**

A measure of the acid level of a substance. The blood pH level shows whether the body is maintaining a normal acid-to-base chemical balance.

- **Pharmacist**

A member of the CF health care team. A pharmacist fills prescriptions and helps manage the use of medicines in the hospital and home. A pharmacist looks to make sure medicines being prescribed do not interact with other medicines and checks the doses to help avoid errors. A pharmacist can show people how to take medicines and tell them what possible side effects to watch for.

- **Phlegm**

Another word for mucus or sputum. See also **mucus**.

- **pH Probe Study**

A procedure to detect gastroesophageal reflux. It measures how much stomach acid is moving into the esophagus from the stomach. A very small tube is passed through the nose down into the esophagus. A pH detector measures the acid level in the esophagus, usually for a 24-hour period. See also **esophagus**.

- **Physical Therapist**

Also called a PT. A health care professional who is trained to help a person make an exercise program to match his or her age, health, and interests. The PT helps people regain strength and endurance through various techniques and improve their mobility and lung function. In some CF centers, the PT will help with airway clearance. See also **lung function**.

- **Physical Therapy**

Treatment by physical means, such as using heat, cold, water, massage, exercise, or mechanical devices. Airway clearance is also a type of physical therapy.

- **Physician**

A doctor who has an MD or DO degree.

- **Physician Assistant**

A health care professional who is trained and licensed to practice medicine under the supervision of a physician. A physician assistant can conduct physical examinations and prescribe medications.

- **Physician Specialist**

A physician (doctor) whose training and practice is in a particular field of medicine or surgery. Typically, physician specialists go on to an internship, residency, and fellowship after medical school. Examples of physician specialists include pulmonologists and gastroenterologists. See also **fellow**.

- **PICC Line**

A type of intravenous (IV) tube that is placed in a vein to deliver fluids and medications. The PICC line has a long tube that goes into a vein in the arm and up into one of the large veins where there is more blood flow. The higher blood flow enables the IV to stay in longer.

- **Pilocarpine Iontophoresis**

A test that collects sweat by stimulating the sweat glands in a small area of skin, usually on the arms or legs. A chemical called pilocarpine is applied together with a small electric current that does not hurt. See also **sweat test**.

- **Pleural Space**

The space between the lung and the chest wall. If there is air in this space, it is called pneumothorax. See also **pneumothorax**.

- **Pneumonia**

An inflammation of the lung tissue usually due to infection. Pneumonia from infection is a serious complication of CF and needs prompt medical treatment.

- **Pneumothorax**

The leaking of air into the space (called the pleural space) between the lung and chest wall. This leak is usually caused by the rupture of an overstretched alveolus (air sac). The lung collapses, either partially or completely. To remove the air, a tube may have to be inserted into the chest. Pneumothorax can be a complication in CF. See also **pleural space**.

- **Polyp(s)**

A small growth of mucous membrane tissue that can cause symptoms. Some people with CF have nasal polyps that can block the nasal passages and sinuses. See also **nasal polyp**.

- **Polypectomy**

A surgical procedure to remove one or more polyps.

- **Portal Hypertension**

High blood pressure in the blood vessels around the liver. Portal hypertension can be seen in cirrhosis (liver disease). See also **cirrhosis**; **liver**; **liver disease**.

- **Positive Expiratory Pressure (PEP)**

A method that gets air into the lungs, behind the mucus, and keeps the airways open. The mucus can then be moved into larger airways where it can be coughed out. PEP is done with a small portable airway clearance device. See also **expiration/expiratory**; **oscillating positive expiratory pressure**; **PEP device**.

- **Postural Drainage and Percussion**

Called PD & P for short. The body is placed in various positions (postures) during percussion. As percussion loosens mucus, the different positions allow gravity to help drain mucus into the large airways. Then the person can clear the mucus from his or her lungs by coughing. A CF health care team member teaches families postural drainage and percussion. See also **airway clearance**; **percussion**; **vibration**.

- **Potassium**

An electrolyte found in the body. Potassium is in many foods, such as oranges, bananas, and potatoes. See also **electrolytes**.

- **Prealbumin**

A type of protein found in the body that can be measured in the blood. This protein is often used as a sign of malnutrition.

- **Prenatal**

A time during pregnancy before the birth of an infant. Mothers get prenatal care to ensure a healthy pregnancy. There are prenatal tests that can be done to diagnose CF in an infant.

- **Prevention**

To take actions to keep a problem from occurring or to avoid an illness or disease complication. With chronic disease, prevention refers to doing things to avoid disease complications or to limit or stop the disease from progressing. Taking a flu vaccine is an example of an action to prevent illness. Doing airway clearance every day is an example of taking action to prevent CF-related lung problems and avoid loss of lung function.

- **Primary Care Physician**

A doctor who serves as the main provider of medical care for a person. The primary care physician is usually expected by medical insurance companies to arrange for referrals to physician specialists and services. See also **physician specialist**; **primary care provider**.

- **Primary Care Provider**

Also referred to as a PCP. Usually a primary care doctor (a family doctor or pediatrician) who sees patients on a regular basis for routine care, such as immunizations and well-child visits; common illnesses or problems, such as ear infections and rashes; and sports or school physicals. A PCP may also be a physician assistant or nurse practitioner. A PCP arranges referrals to specialists. See also **nurse practitioner**; **pediatrician**; **pediatric nurse practitioner**; **physician assistant**.

- **Productive Cough**

A cough that brings up sputum or mucus from the lungs. This type of cough is common in people with CF. It is also described as a “wet” or “loose” cough.

- **Prognosis**

A prediction of the course or effect of a disease. A way of describing the chance of a person recovering from a disease or how long a person is likely to live. The prognosis for different people with the same disease can vary greatly, and the prognosis of one person may change over time.

- **Progression**

A change over time in a disease—most often a worsening or increase in disease and its related problems. In CF, lung disease can get worse over time and there can be a progressive loss of lung function. See also **lung function**.

- **Protease**

One of the enzymes produced in the pancreas. Protease helps break down proteins in food in the intestine. Protease is included in pancreatic enzyme supplements in CF. See also **enzymes**; **pancreatic enzymes**.

- **Protein**

A type of nutrient found in the diet that is important to all cells in the body.

- **Pseudomonas**

A group of bacteria. In CF, *Pseudomonas* bacteria can cause lung infections.

- **Pseudomonas Aeruginosa**

Also called *P. aeruginosa* for short. A type of bacteria that, in CF, is most commonly found in airways. In CF, *Pseudomonas aeruginosa* is the major bacterial cause of lower respiratory tract infections.

- **Psychiatrist**

A doctor with special training to diagnose and treat people who have mental or emotional health issues. A psychiatrist may often prescribe medications, and sometimes may also prescribe other therapies. At times, a psychologist and psychiatrist may work together with patients and families. See also **diagnose**.

- **Psychologist**

A health care professional with expertise in assessing and treating problems with behavior, learning, emotions, and group/family interactions. A psychologist helps with problems, such as depression, learning disabilities, or behavior problems. A CF team psychologist may also focus on the problems people may have managing CF. These include balancing treatments and other aspects of life, helping those who are having trouble developing self-management skills, and helping children and teens manage peer issues related to CF. Psychologists do not prescribe medications.

- **Puberty**

The period in which a child becomes capable of sexual reproduction (becomes fertile). This is also a period of increased growth during adolescence, or the teenage years. See also **adolescence**; **fertile**.

- **Puffer**

Another name for an inhaler. See **metered dose inhaler**.

- **Pulmonary**

Referring to the lungs.

- **Pulmonary Function Tests**

Also called PFTs for short. Tests that show how well a person can breathe. The tests measure how fast air moves and the volume of air moving in and out of the lungs. The tests can also measure how the airways respond to inhaled medications.

- **Pulmonary Rehabilitation**

A physical therapy program that helps a person with lung problems exercise and return to the highest level of activity possible. It helps people with CF improve their mobility and lung function. See also **physical therapist**; **physical therapy**.

- **Pulmonologist**

A physician (doctor) who has special training in diseases of the lungs and breathing problems. Some specialize in the care of children and teens, and others specialize in the care of adults.

- **Pulse Oximeter**

A machine that measures oxygen saturation in the blood. The amount of oxygen in the blood is measured by placing a light probe over a finger or a toe. See also **oximetry/pulse oximetry; oxygen saturation**.

Q

- **Quality of Life**

A person's satisfaction or happiness with different aspects of his or her life, such as work, relationships with others, and physical health.

R

- **Rale**

See **crackle(s)**.

- **Reactive Airway Disease**

Also called RAD for short. See **asthma**.

- **Rectal Prolapse**

When the inner lining of the rectum (the last part of the large intestine) comes out (prolapses) through the anus. In CF, this may be caused by straining to pass a large or hard stool from the poor digestion of food. Rectal prolapse usually is prevented with good control of malabsorption and malnutrition. See also **malabsorption; malnutrition**.

- **Rectal Temperature**

The most accurate measure of an infant's or young child's temperature. A rectal thermometer is placed in the rectum. See also **rectum**.

- **Rectum**

The last part of the large intestine, ending at the anus. See also **anus**.

- **Renal**

Relating to the kidneys.

- **Replacement Enzymes**

See **pancreatic enzymes**.

- **Research**

Through research, scientists better understand how the body works, what happens in disease,

and ways to treat and prevent disease.

In **basic science research**, scientists seek to increase basic knowledge. For example, they try to understand how cells work and see what is different in CF. One type of basic science research is how genes affect body functions.

In **applied or translational research**, scientists take the findings of basic science research and use them to solve problems of everyday life. They might, for example, look for ways to change the CF gene to reduce the problems in CF, or how to design equipment to deliver medicine to the lungs.

In **clinical research**, scientists seek new and better ways to diagnose and treat patients who have a disease. In CF, they might look for ways to diagnose CF earlier, or study new antibiotics to treat infection.

- **Resident**

A resident is a physician (doctor) who has graduated from medical school and is in training to become a particular type of doctor, such as a pediatrician. A first-year resident is sometimes called an intern. Residents provide medical care to patients in teaching hospitals and clinics under the supervision of an attending physician.

- **Resistance**

The ability to remain unaffected by something that might be harmful. Vaccines, for instance, are given to help the body create resistance against certain viruses or bacteria that may cause disease. Sometimes bacteria become resistant to an antibiotic. This means the antibiotic can no longer kill or limit the growth of the resistant bacteria.

- **Resistance Exercises**

Exercises that are done to build and strengthen muscles, such as lifting weights, sit-ups, pull-ups, and using resistance bands. These may be done by themselves or as part of a warm-up with aerobic exercise. See also **aerobic**.

- **Respiration**

Breathing. How the body takes in and uses oxygen from the air and gets rid of carbon dioxide.

- **Respiratory Infections**

See **lower respiratory tract infections; lung infection(s)**.

- **Respiratory Rate**

The number of breaths a person takes in 1 minute.

- **Respiratory System**

The parts of the body used for breathing. The upper respiratory tract includes the nose, sinuses, mouth, and throat. In the lower respiratory tract are the trachea (windpipe), lungs, and the airways below the vocal cords. See also **alveoli**; **bronchi**; **bronchioles**; **diaphragm**; **lung tissue**.

- **Respiratory Therapist**

RT for short. A member of the CF health care team. This health care professional is also called a respiratory care practitioner (RCP for short). An RT teaches airway clearance, and the correct use of inhaled medicines, oxygen therapy, or other breathing support machines.

- **Respiratory Treatments**

See **breathing treatments**.

- **Retractions**

Retractions are the sucking in of the muscles between the ribs with breathing. Retractions can also be seen below the rib cage and in the neck just above the sternum (breastbone) when a child is working hard to breathe. Caused by the use of breathing muscles when it is hard to move air in the lungs.

- **Reward**

A reward (often called a reinforcer) is anything, such as an activity, object, or praise, that a person (parent) gives to another (child) to “reinforce” or increase the chance that a certain behavior will occur again in a similar situation.

- **Rhinovirus**

A common type of virus that can cause upper and lower respiratory tract infections (from the common cold to pneumonia) in people of all ages. It is the most frequent cause of the common cold.

- **RSV**

Short for respiratory syncytial virus. A common type of virus that causes upper and lower respiratory tract infections from the common cold to pneumonia. In young children it is the major cause of pneumonia and bronchiolitis. See also **bronchiolitis**; **pneumonia**.

S

- **Saline**

A mixture of salt and water. Normal saline is made with a level of salt similar to the body’s own fluids. Hypertonic saline has a higher salt content than does body fluid. Saline may be used as an IV fluid and in breathing treatments. See also **hypertonic saline**.

- **Secondhand Smoke Exposure**

When those who do not smoke breathe in air polluted with tobacco smoke from other people’s cigarettes, pipes, or cigars. Children with CF are at greater risk for respiratory problems, such as pneumonia, bronchitis, or reduced lung function if they are exposed to tobacco smoke. Eliminating a child’s exposure to tobacco smoke can help prevent worsening of CF.

- **Sedated/Sedation**

A person is given medication to become more calm and/or go to sleep. The person is breathing on his own when sedated. Sedation may be used for some tests or procedures.

- **Self-Management**

How people with chronic disease manage their health and the disease. Self-management includes watching for possible problems or changes in symptoms, working with the health care team to make a plan to solve the problems, and using prescribed medications and therapies to prevent and treat disease problems or complications. The CF Family Education Program (CF FEP) helps parents and children with CF learn how to be good CF managers. See also **CF manager**.

- **Semen**

Also called seminal fluid. A thick, white fluid that is produced in the male sex organs and leaves the body through the penis during ejaculation. Sperm normally mixes with semen. In CF, a male can have semen without any sperm. See also **ejaculation**; **sperm**.

- **Seminal Vesicles**

Glands in the male reproductive system that help form semen. See also **semen**.

- **Sensitivity**

A test that is done to see what antibiotics work to kill or limit the growth of specific bacteria. In CF, this is usually done on a sputum culture. Bacteria that do not grow well when exposed to an antibiotic are said to be sensitive to that antibiotic. This test helps doctors choose the best treatment for a bacterial infection. See also **antibiotics; bacteria**.

- **Sexual Intercourse**

The act of a male's penis being inserted into a female's vagina. Semen is ejaculated from the penis at times during intercourse. If sperm is in the semen and connects with a female's egg, conception can occur. See also **conception; ejaculated; penis; sperm**.

- **Sexually Transmitted Diseases (STDs)**

Also called sexually transmitted infections (STIs). Diseases that can be spread to another person through sexual contact, such as AIDS (HIV virus), gonorrhea, and genital herpes virus.

- **Shaping**

Shaping is a way to help a person learn complex behavior in smaller steps and over a period of time (sometimes months or years). For example, to help a child self-manage respiratory care, a parent would first expect and praise a toddler for cooperating with breathing treatments. As the child gets older, the parent would reinforce the child for sometimes reminding the parent that it is time for the treatments. Later, the parent would praise the child for setting up and taking the treatments on her own. Finally, the child is able to do it all, including being responsible for cleaning the equipment, making sure prescriptions are filled, noticing when equipment is worn, and requesting new equipment when needed.

- **Sibling or Sib**

A brother or sister.

- **Side Effects**

Unwanted reactions to medications or treatments. Some side effects can be avoided by taking a medicine in a specific way or adjusting the dose. Some side effects can be very serious and may require a person to stop the treatment. Sometimes a person may need to tolerate mild side effects in order to get the benefit of a medicine if there is not a better choice. Parents should not stop or adjust their child's medications on their own, but should instead let their health care team know if they are concerned about any side effects.

- **Signs**

The clues of an illness or problem that can be observed by another person. Fever may be a sign of infection. Doctors listen to the lungs for wheezing, a sign of a lung problem. See also **symptoms**.

- **Sinus(es)**

The sinuses are hollow spaces in the skull found under the forehead, in the cheeks, and around the nose. The sinuses are lined with the same type of cells that are found in the airways. The sinuses help warm the air a person breathes in and affect how his or her voice sounds. A person can have problems with congestion in his or her sinuses because of allergy or infection. People with CF usually have sinuses filled with abnormal mucus (similar to the mucus in the airways) and can have chronic sinus infections as well.

- **Sinusitis**

An inflammation of the sinuses usually due to infection or allergy. Sinusitis from an infection is a complication of CF. See also **sinus(es)**.

- **Small Intestine**

See **intestines (small and large)**.

- **Social Worker**

A health care professional who helps families adapt to chronic disease care and cope with family and social problems related to health problems. The social worker helps families deal with obstacles that may interfere with their ability to handle health problems. The social worker also helps people figure out how to access help with health care insurance, career choices, and school issues.

- **Sodium**

An electrolyte found in the body. Sodium and chloride make salt. See also **chloride; electrolytes**.

- **Spacer**

See **holding chamber**.

- **Specialist**

Also referred to as a subspecialist. A physician or other health care professional (such as a physical or respiratory therapist) who has training and practice in a particular field of medicine or surgery.

- **Sperm**

The cell that is made by a male and combines with a female's egg for conception. See also **conception**.

- **Spirometry**

A pulmonary function test that measures how much air moves in and out of the lungs and how fast the air can flow. Spirometry can be done regularly to check how the lungs are doing. Often when an infection is present, the spirometry results will be worse and then will improve with successful treatment. The CF doctor will look at results from several measures, including FEV₁ and FVC. See also **FEV₁; FVC; pulmonary function tests**.

- **Spleen**

An organ in the left upper part of the abdomen near the stomach. The spleen stores blood and filters out old blood cells and other substances, and helps the body fight certain infections. It can be enlarged with portal hypertension. See also **abdomen; portal hypertension**.

- **Sputum**

Another name for mucus or phlegm coughed up from the lungs. See also **mucus**.

- **Sputum Culture**

A test done on mucus coughed up from the lungs. See also **cultures**.

- **Staphylococcus Aureus**

Also called *S. aureus*, or “staph” for short. A type of bacteria often found on the skin and sometimes in the airways of people with CF. *Staphylococcus aureus* can cause infection. Methicillin-resistant *Staphylococcus aureus* or MRSA is one type of staph that is resistant to many antibiotics. See also **antibiotics; bacteria; methicillin-resistant Staphylococcus aureus; resistance**.

- **Steatorrhea**

Fat in the stool.

- **Stenotrophomonas Maltophilia**

A type of bacteria. In CF, *Stenotrophomonas maltophilia* is sometimes found in the airways and may cause lung infection.

- **Streptococcus Pneumoniae**

A type of bacteria that may cause infections, such as ear infections and pneumonia. These bacteria can grow in the airways of a person with CF and can cause bronchitis. Infection from some strains of *Streptococcus pneumoniae* can now be prevented with a vaccine. (Some people call this the “pneumonia vaccine.”)

- **Supplements**

Nutrients that can be taken or mixed with other foods to add extra calories or protein to the diet. See also **nutrients**.

- **Surgeon**

A physician (or doctor) who does procedures and operations, such as surgery for meconium ileus, or gastrostomy tube placement. See also **gastrostomy tube; meconium ileus**.

- **Sweat Glands**

Sweat glands are clusters of cells found in the skin that secrete (produce and release) sweat. See also **exocrine glands; sweat test**.

- **Sweat Test**

A test used to diagnose CF. The sweat glands are exocrine glands, and in CF they produce sweat that is saltier than normal. Salty sweat has high levels of sodium and chloride. A high chloride level usually means the person has CF. See also **chloride; exocrine glands; pilocarpine iontophoresis; sodium**.

- **Symptoms**

The things people notice when they are ill or have other problems. A person with CF may feel tired or have chest pain with an infection. Stomach cramps may be a symptom of malabsorption. See also **signs**.

- **Synergy Effect**

When two things work together to have a stronger effect than each would alone. For example, there are combinations of antibiotics that may kill or slow bacteria when used together. See also **antibiotics; bacteria**.

T

- **Target Heart Rate**

The speed at which a person’s heart should beat during exercise to reach a good aerobic effort. The CF doctor or physical therapist helps people figure out their target heart rate for exercise. See also **aerobic; physical therapist**.

- **Teeth Enamel**

The hard coating covering teeth made up of minerals and protein.

- **Testes**

The round organs in the scrotum of the male where hormones (testosterone) and sperm are made. See also **sperm; testosterone**.

- **Testosterone**

A male sex hormone that comes from the testes. Testosterone is important in male sexual development and function. See also **hormone**; **testes**.

- **Therapies**

Medicines or other treatments that are prescribed to help control or resolve an acute or chronic health problem. Therapies in CF include medicines, airway clearance, and a high-calorie diet.

- **Think and Act**

Two of the actions people with CF and families do day to day as they manage a chronic disease. The first step is to **Watch and Discover** a change or problem. Next, to **Think** about possible solutions, set goals, and make a plan. Then, choose a solution and **Act**. After taking action, a person watches to see if the action is successful or if a different solution needs to be tried. The CF health care team helps people with CF learn how to **Think and Act** to manage CF. See also **CF manager**; **Watch and Discover**.

- **Thrush**

An infection in the mouth or throat caused by the fungus, *Candida albicans*. White patches or ulcers appear on the mouth, tongue, and throat. See also **fungus**.

- **Tissue**

A group of cells of a similar type, having a similar function (for example, lung tissue).

- **Total Parenteral Nutrition (TPN)**

Nutrients are given through a tube put into a large vein (central line). This lets the body get nutrients when it is not possible to get them through the digestive system. The solution used most often contains dextrose (a form of sugar), amino acids (simple proteins), lipids (fats), electrolytes, minerals, and vitamins.

- **Trachea**

The windpipe that connects a person's lungs to the throat. See also **lower respiratory tract**; **respiratory system**.

- **Trough Level**

The lowest level of a medicine in the bloodstream as measured by a drug level test. The body needs to break down and clear a medicine between doses. If it cannot clear the medicine, the level may build up and cause side effects. The level is drawn just before the next dose is given. Trough levels are measured to make sure the right dose is being given to work safely and well. See also **side effects**.

U

- **Upper GI**

Also called a barium swallow. An X-ray that is done to look at the esophagus, stomach, and small intestine. A person is asked to swallow barium, a substance that can be seen on an X-ray. Often this test is done to look for signs of gastroesophageal reflux. See also **esophagus**; **gastroesophageal reflux**.

- **Upper Respiratory Tract**

Includes the nose, sinuses, mouth, and throat.

- **Upper Respiratory Tract Infections**

Also called URI or URTI for short. An infection in the upper respiratory tract above the vocal cords including the nasal passages, sinuses, and throat. URIs may be caused by viruses or bacteria. The common cold, for example, is a URI that may be caused by many viruses. See also **lower respiratory tract infections**.

- **Urinalysis**

A test that looks at the urine for certain chemicals and cells. Can help detect a urinary tract infection or kidney problems. In uncontrolled diabetes, there may be glucose in the urine.

- **Urinate**

To empty the bladder of urine. Commonly called peeing or passing urine.

- **Urine**

A liquid waste made by the kidneys that leaves the body. Sometimes called “pee.”

- **Uterus**

A muscle, also called the womb, where the baby grows until birth. Part of the female reproductive system.

V

- **Vaccination**

Giving a person a vaccine containing material that comes from a bacteria or virus (not the whole live germ) to help the body build immunity against infection with the bacteria or virus. See also **immunizations**.

- **Vaccine**

See **immunizations**; **vaccination**.

- **Vagina**

The canal that leads from the uterus to the outside of the body. Part of the woman's reproductive system.

- **Vas Deferens**

The tube that carries sperm from the testes to the penis. Males with CF usually do not have a vas deferens, which results in infertility. See also **congenital bilateral absence of the vas deferens; infertility**.

- **Vial(s)**

Small plastic containers with twist-off tops containing liquid medicine. The single dose of medicine is poured into the nebulizer cup for breathing treatments.

- **Vibration**

A technique used with postural drainage and percussion to help loosen and remove mucus from the lungs. The hands are used to make rapid vibrating motions on the chest wall as the person breathes out. A CF health care team member teaches families how to do vibration. See also **postural drainage and percussion**.

- **Viral**

Caused by or related to a virus or viruses. See **viruses**.

- **Viruses**

Organisms smaller than bacteria. Many different viruses cause infection. There are no medications to treat most viruses. Viral illnesses include chickenpox, the common cold, RSV, and influenza (flu). See also **RSV; viral**.

- **Vital Signs**

Measures of body functions including temperature, heart rate (pulse), respiratory rate, and blood pressure.

- **Vitamins**

Chemicals found in the diet that are important to the body. There are a number of vitamins that the body uses. Some vitamins are fat-soluble and the body needs enzymes to absorb them. A person with CF and pancreatic insufficiency needs extra amounts of the fat-soluble vitamins (A, D, E, and K).

W

- **Watch and Discover**

Two of the actions people with CF and their families need to do day to day as they manage a chronic disease. First, they Watch for new or changing symptoms. Then, they Discover what possible causes there are for the changes. Next, they Watch to see if the plan to handle a problem is working. The CF health care team helps families and people with CF learn how to Watch and Discover problems in CF. See also **symptoms; Think and Act**.

- **Water-Soluble**

Able to be dissolved in water. Water-soluble vitamins, such as vitamin C, are easier to absorb than fat-soluble vitamins.

- **Wheezing**

A high-pitched whistling sound heard in the lungs. It may be heard with a stethoscope, or sometimes out loud without a stethoscope. The sound is made as air moves through narrowed airways. Wheezing occurs with airway obstruction from bronchospasm or mucus. See also **bronchospasm**.

- **Windpipe**

See **trachea**.

X

- **X-Ray**

Short for radiograph. A test that shows images of the inside of the body (for example, the lungs or bones of the hand).

Z

- **Zinc**

A mineral in the diet that is important to the body. Low levels can occur at times in CF.

