

CYSTIC FIBROSIS FOUNDATION
ADDING TOMORROWS

An Introduction to Cystic Fibrosis

For Patients and Their Families

SIXTH EDITION

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DEDICATION

This book is dedicated, with love and respect, to people with CF and their families, who teach us daily about life and CF.

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How to Use This Book

If you are the parent or relative of someone who has just been diagnosed with cystic fibrosis (CF) or an adult who has just learned that you have CF, you may be worried, confused or scared. You might have questions about CF and its treatment. This is normal and expected. This book will help you learn more about CF. It will answer questions, clear up confusion and tell you how you can learn more and where to find support.

This book has 15 chapters. Many chapters end with "Review Questions" so you can check your learning. As you read, it may be helpful to write down your own questions in the space given. Cover one chapter at a time. Bring this book and your questions to meetings with your CF care team at your CF care center to get answers. We are always learning more about CF and how to treat it. This book cannot answer all of your questions, but it will help you better understand CF and its treatment.

All of the words that are bolded and italicized at their first mention in this book are in "Appendix A: Medical Words to Know" at the back of the book. The "Resource List" in Appendix B can direct you to additional information about many of the topics covered in this book. Appendix C can help you learn more about postural drainage and percussion, an airway clearance technique. Appendix D has examples of blank forms you can copy and use to help you organize your or your child's CF care. Cystic Fibrosis Foundation–accredited care centers are listed in Appendix E.

You may have been given a DVD to go with this book. It will introduce you to CF and the CF care team. This book will cover the same topics, but in more detail. Keep both the book and DVD for future use, or to share with relatives and friends who want to learn more about CF.

PART I

Definition and Diagnosis

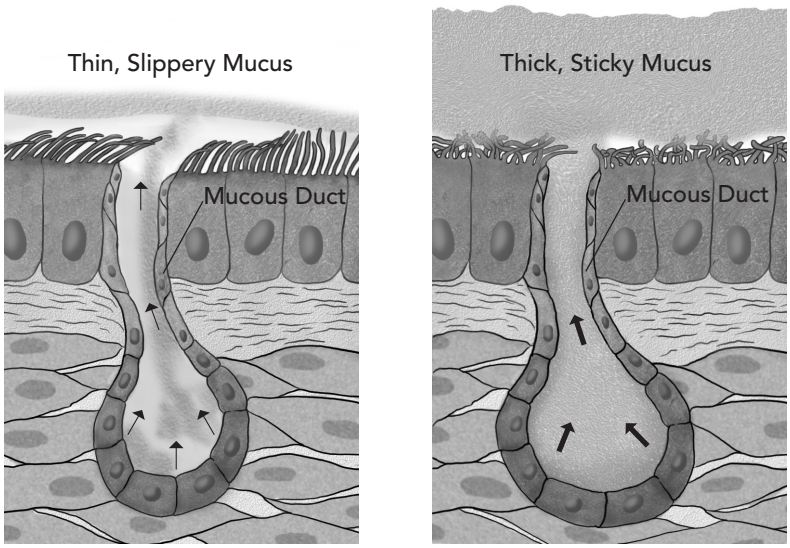


What Is Cystic Fibrosis?

Cystic fibrosis (usually called CF) is an ***inherited**** disease. It causes certain glands in the body to not work properly. These glands are called the exocrine (outward-secreting) glands.

Exocrine glands normally make thin, slippery ***secretions*** including sweat, ***mucus***, tears, saliva and digestive juices. These secretions move through ***ducts*** (small tubes) to the surface of the body or into hollow organs, such as ***intestines*** or ***airways***. Exocrine glands and their secretions help the body function normally.

Lung Cells



Non-CF
In a mucous gland, secretions easily move through the duct.

CF
CF causes the mucus to be thick and sticky.

In CF, exocrine glands (except sweat glands) make mucus that is too thick and sticky. This mucus plugs ducts and other passageways. ***Mucous plugs*** are most often in the lungs and intestines and can cause problems with breathing and digestion.

CF also affects sweat glands. The levels of salt (sodium and chloride) and potassium in the sweat are too high. This may cause problems during times of increased sweating.

CF does not affect ***endocrine glands***. Endocrine glands and exocrine glands are different. Endocrine (inward-secreting) glands make ***hormones*** that pass into the blood.

**Bold and italicized words are defined in Appendix A, "Words to Know," page 159.*

What CF Is Not

Many people are confused about CF. Below are some common topics that may cause confusion:

CF Is Not Contagious.

Because CF causes ***coughing***, some people think you can "catch" it. CF is ***genetic***. You are born with it. No one can "catch" CF. You cannot give it to anyone else. We will talk more about how a person gets CF later.

CF Is Not Caused by Anything the Mother or Father Did, or Did Not Do, Before or During Pregnancy.

Parents feel responsible for what happens to their children. When their children have CF, some parents feel guilty. Nothing parents did before or during pregnancy caused CF!

CF Does Not Affect the Brain.

Some people confuse CF with cerebral palsy or CP. CF does not affect the brain, ***nervous system*** or the capacity to learn.

CF Has No Cure at This Time.

But with treatment, most people with CF grow up and lead active, full lives. A lot of time, energy and money are being spent to find new, better ways to treat CF.



CF does not affect the brain. Children with CF have the same ability to learn as children without the disease.



About 30,000 people in the United States have CF.

Is CF a New Disease?

CF is not a new disease. As early as 1595, writings suggested that there were children who likely had CF. "Cystic fibrosis of the ***pancreas***" was first described in 1936 by Dr. Guido Fanconi of Switzerland. Dr. Dorothy Andersen at Babies and Children's Hospital in New York City wrote the first full report of CF as its own disease in 1938. We have learned much more about CF since then.

How Common Is CF?

CF is one of the most common genetic (inherited, ***hereditary***) disorders in white people. In the United States, CF occurs in one of every 3,200 live white births. It occurs in one of every 15,000 live African American births. CF is uncommon in Asians and most Native American tribes. It is being seen more often in Hispanics. It occurs equally in males and females.

In the United States, about 800 to 900 persons are diagnosed with CF yearly. There are approximately 30,000 people with CF in the United States today. That number is rising as more people are diagnosed, treated sooner and living longer.

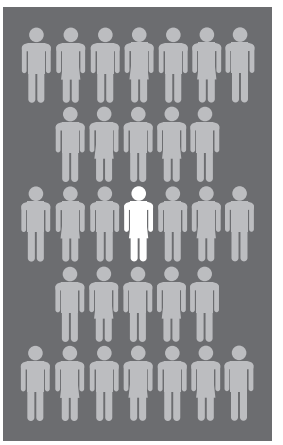
Genes, Chromosones and How a Person Gets CF

In the United States, one person in every 31 carries a ***mutation*** of the CF ***gene***. That means more than 10 million Americans have one copy of a CF gene mutation. People who carry one CF gene mutation do not have CF. But not all gene ***carriers*** can be identified. ***Research*** to develop a test for all carriers is ongoing.

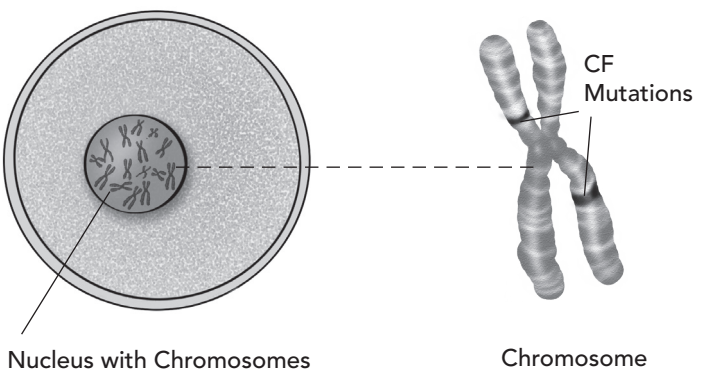
Every person with CF was born with CF. It is a genetic disease that starts at ***conception***. The age when ***symptoms*** start, type of symptoms and the severity of the disease vary. In most people, the lungs are the most affected part of the body; in others, the ***digestive system*** may be affected the most.

Conception occurs when the mother's ***egg*** joins the father's ***sperm***. Both the egg and the sperm have thousands of genes. Genes are the main units of heredity. They decide body traits including eye and hair color, height, facial features and many health conditions. All people probably have seven or eight genes that could be connected with serious health problems such as CF.

Genes are found on ***chromosomes***. These are threadlike structures found in the ***nucleus*** of all ***cells***. Each chromosome has thousands of different genes. Genes come in pairs. Each parent gives one gene from his or her pair to make up a new pair for the child. Genes pass on family traits from one generation to the next.



One in every 31 Americans is a symptomless carrier of one copy of the gene that causes CF.



Genes (the dark lines on the chromosome) are located on chromosomes. They determine things such as eye color, hair color, height and CF.

An Autosomal-Recessive Disease

CF is an ***autosomal-recessive*** disease. Autosomal means that the CF gene is not on the sex chromosome. In other words, both males and females can get CF. If the CF gene mutation is paired with a normal gene, the normal gene will do all the work and the CF gene mutation will be recessive. This person will not have CF but will be a carrier. A carrier is someone with one CF gene mutation and one normal gene. A carrier has no symptoms and no disease.

Both parents of a child with CF are carriers of at least one CF gene mutation. They inherited this gene from one of their parents, who inherited the gene from one of their parents.

Every child with CF got genes for this disease from both the mother and father. So a CF gene mutation is present on both sides of the family.

When both parents are carriers and each gives the child a CF gene mutation, there is no gene that works right. The child will have CF.

More about the CF Gene

The gene that causes CF was identified in 1989. Genes are the main units of heredity. Genes have small building blocks called ***base pairs***. The gene that causes CF has a mutation or change in the genetic material, causing an exchange or loss of a building block. Since 1989, more than 1,500 different mutations of the CF gene have been found, and new ones are still being found. Scientists are studying the effects of the different CF gene mutations.

Finding the CF gene has helped us learn how and why this gene causes CF. That information has led to research on new ways to treat CF. Learn more about "The Future and CF Research" in Chapter 15.



If both parents are carriers of the CF gene mutations, there are tests that can be done ***prenatally*** *to see if the* ***fetus*** *has CF.*

Can CF Carriers Be Identified?

Because a carrier has no CF symptoms, we often do not know if the person is a carrier until he or she becomes the parent of a child with CF. Carriers of a CF gene mutation often can be identified if a relative has CF. First, the blood or cells brushed from the inside of the cheek of the person with CF are tested. If the mutations in the CF gene are identified, then relatives can be tested for those same mutations to see if they are carriers.

Finding CF carriers in the general public is harder. Known gene mutations can be looked for, but we cannot know if the person is a carrier of an unknown mutation. Also, most laboratories only

test for the most common mutations. Screening for all mutations is difficult. So screening the general public for carriers of the CF gene mutation is not possible.

What about the Risk of CF in Future Pregnancies?

When both parents carry a CF gene mutation, their children will not always have CF. The CF gene is carried by half of the father's sperm and half of the mother's eggs.

Inheriting CF

The father's sperm with a CF gene mutation may join with the mother's egg that has a CF gene mutation. With two CF gene mutations, the child has CF.

A sperm with a normal CF gene may join with an egg that has a CF gene mutation. The child will not have CF but will be a carrier.

A sperm with the CF gene mutation may join with an egg with a normal CF gene. The child will not have CF but will be a carrier.

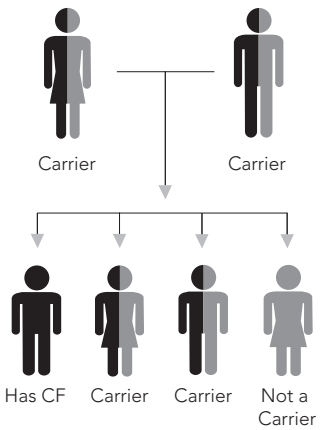
A sperm with a normal CF gene may join with an egg with a normal CF gene. The child will not be a carrier and will not have CF.

The chart on this page shows what might happen when the sperm and egg of parents who carry a CF gene mutation join.

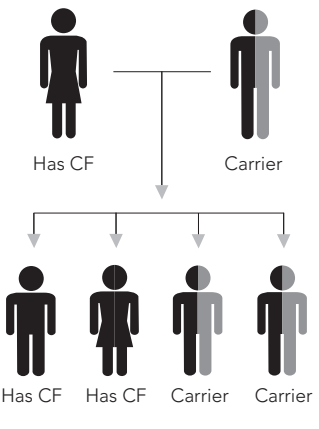
How a Person Gets CF

To have CF, you must get one copy of the CF gene from each parent. That means that each parent must be a carrier of the CF gene.

When two people who are carriers have a child, there is a 25 percent chance of having a child with CF.



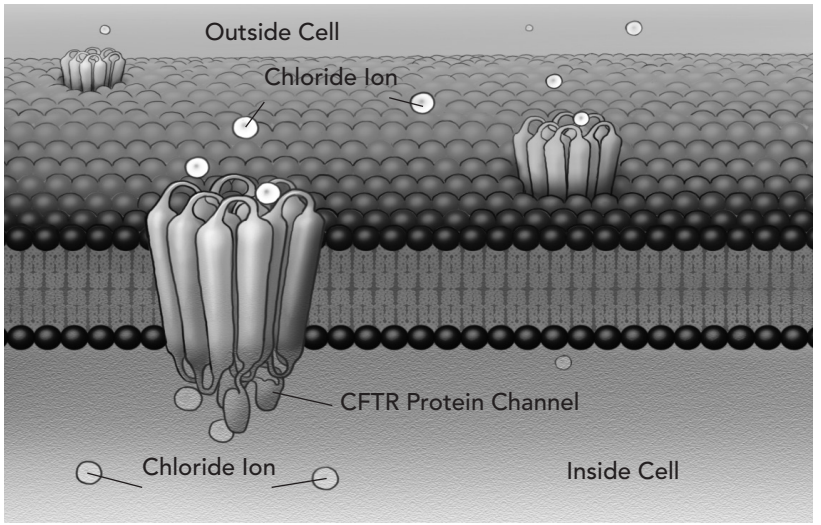
When one parent has CF and one parent is a carrier, there is a 50 percent chance having a child with CF.



When both parents carry a CF gene mutation, **each baby** has a:

- One in four (25 percent) chance of not being a carrier and not having CF
- Two in four (50 percent) chance of being a carrier but not having CF
- One in four (25 percent) chance of having CF

Each pregnancy has these odds, even if the parents already have a child with CF.



Chloride Ions move from inside to outside a cell through the CFTR protein channel.

How Do Altered Genes Cause CF?

CF is genetic. The healthy version of the gene directs the making of a protein called the ***Cystic Fibrosis Transmembrane conductance Regulator (CFTR)***. This protein forms a channel so that salt (chloride and sodium) and water can move in and out of cells (called ***ion transport***). The CF gene mutation changes CFTR so that the protein does not work properly. As a result, salt and water transport breaks down (abnormal ion transport). The biggest changes are in the sweat and mucous glands. Knowing how CFTR works gives clues to researchers about new potential treatments.

Sweat Glands

The CF gene mutation causes a chain of events that leads to abnormal secretions. In CF, the sweat glands make sweat that is more salty than in someone who does not have CF. Research shows that abnormal salt transport in cells occurs in all of the exocrine glands in CF. This problem is called the ***basic defect*** in CF.

Mucous Glands

Normal mucus is thin and slippery. In the airways, it helps remove dust and ***germs***. It also lubricates the ducts and passageways of other organs, like oil in a car.

In CF, the changes in salt transport cause changes in water transport. The mucus loses some of its water and becomes thick and sticky. This thick mucus blocks tubes and organ passageways.

Mucous blockages cause many CF symptoms. The most serious are:

Chronic Lung Disease

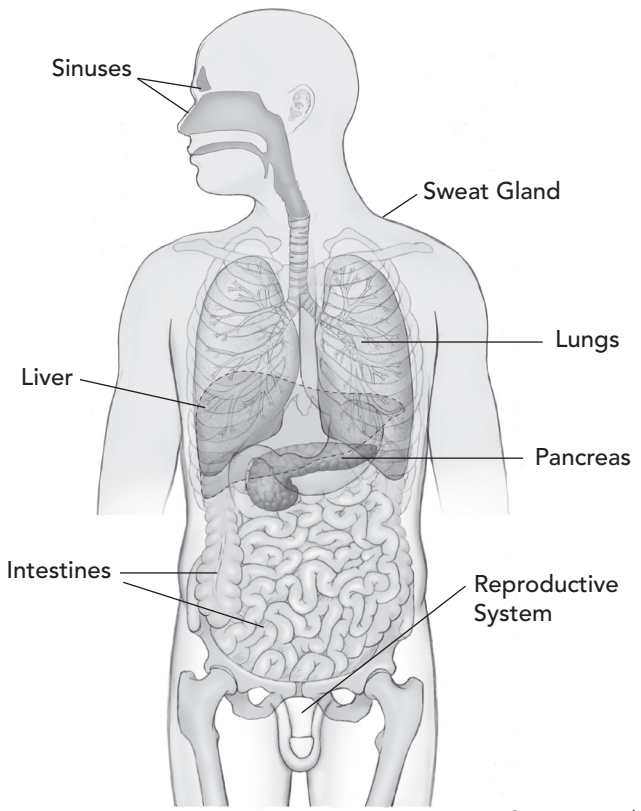
Lung airways clogged by thick mucus can get infected and inflamed. Infections and ***inflammation*** cause ***chronic*** lung disease. At some point, all people with CF get chronic lung disease.

Impaired Digestion

Digestion problems occur in 90 percent of people with CF. Thick mucus keeps digestive ***enzymes*** from the pancreas from getting to the intestines where they are needed to help digest food.

CF also affects other body parts including the liver and ***reproductive system***. For most people with CF, these problems are not as bad as the effects on breathing and digestion. Later chapters will look at the effects on the lungs, intestines and other body parts.

We do not know everything about CF, but we have made great strides. Scientists still study CF gene mutations, the CF gene's protein product and the basic defect to help treat the cause of CF, not just treat the symptoms.



CF causes changes in many parts of the body, including the lungs, pancreas, liver, intestines, reproductive system and sweat glands.

Review Questions

Answers follow the questions. More than one answer may be correct.

1. CF is a disease of the:
   a. Blood
   b. Brain
   c. Exocrine glands
   d. Endocrine glands
2. A person gets CF by:
   a. Being near someone with CF who coughs
   b. Touching a dirty tissue
   c. Inheriting two CF gene mutations
   d. Touching a person who has CF
3. Exocrine glands produce these:
   a. Mucus
   b. Sweat
   c. Tears
   d. Saliva
   e. Hormones
   f. Digestive juices
4. CF affects these parts of the body:
   a. Liver
   b. Brain
   c. Lungs
   d. Pancreas
   e. Intestines
5. Which of these is a symptom of carriers of a CF gene mutation?
   a. Cough
   b. Wheezing
   c. Pneumonia
   d. Abnormal stools
   e. Difficulty gaining weight
   f. All of the above
   g. None of the above
6. When parents already have a child with CF, what are the odds that their next child will have CF?
   a. The same odds apply to every pregnancy.
   b. There is no chance the next child will have CF.
   c. The parents will be less likely to have another child with CF.
   d. The parents will be more likely to have another child with CF.
7. In CF, mucus and secretions become too:
   a. Thin and dry
   b. Thin and watery
   c. Thick and sticky
8. Which statement about CF is true?
   a. Smoking during pregnancy increases the chance of having a child with CF.
   b. CF is not caused by anything the parents did or did not do before or during the pregnancy.
   c. Eating the right foods during pregnancy reduces the chance of having a child with CF.
   d. The chance of having a child with CF is decreased by not drinking alcohol during pregnancy.

9. When does CF begin in a person?
   a. At conception
   b. At about 2 years of age
   c. At different times for different people
   d. When the person first begins to feel sick

10. Which of these statements about CF genes is true?
   a. One in every 31 people in the United States carries a CF gene mutation.
   b. Both parents of a child with CF carry a CF gene mutation.
   c. If the mother or father carries a CF gene mutation, the child will have CF.
   d. A child must inherit one CF gene mutation from each parent to have CF.

11. Which of the following statements is true?
   a. Anyone can be tested to see if they carry a CF gene mutation with 100 percent accuracy.
   b. A family member of a person with CF can be tested for carrier status with a high level of accuracy.

Answers

To learn more, turn to the page number shown after the answer.

1. C
CF is a disease of the **exocrine glands**. Page 11.

2. C
A person gets CF by **inheriting two CF gene mutations**. Page 15.

3. A, B, C, D, F
Exocrine glands produce **mucus, tears, sweat, saliva and digestive juices** produced in the pancreas. Hormones are made by the endocrine glands. Page 11.

4. A, C, D, E
CF affects the **liver, lungs, pancreas and intestines**. CF does not affect the brain. Page 12, 17.

5. G
None of the above is a symptom of carriers of a CF gene mutation. Carriers of CF have **no symptoms of CF** and do not have CF. Page 14.

6. A
The same odds apply to every pregnancy. Because they already have a child with CF, both parents are carriers of one CF gene mutation. Therefore, they have a one-in-four chance of having a child with CF from each and every pregnancy. Page 15.

7. C
In CF, mucus and secretions become too **thick and sticky**. Page 11.

8. B
It is true that **CF is not caused by anything the parents did or did not do before or during the pregnancy**. Page 12.

9. A
CF is inherited and begins at the moment of **conception**. The age when symptoms first appear varies; they can appear at birth or anytime during a person's life. Page 13.

10. A, B, D
It is true that **one in every 31 people in the United States carries a CF gene mutation, that both parents of a child with CF carry a CF gene mutation and that a child must inherit one CF gene from each parent to have CF**. It is not true that if the mother or father carries a CF gene mutation, the baby will automatically have CF. Each parent gives one gene to make up the child's gene pairs. Both parents must be carriers and each give the child a CF gene mutation for their child to have CF. Page 14-15.

11. B

It is **true** that a family member of a person with CF can be tested for carrier status. In all likelihood, the CF gene mutation will be the same for all members of the family. Because there are more than 1,500 CF gene mutations, a person cannot be tested with 100 percent accuracy. Page 14.

Notes and Questions

Chapter 2: How CF Is Diagnosed

Diagnosing CF

Cystic fibrosis (CF) is ***diagnosed*** by using a medical history, physical examination and laboratory tests.

Medical History

The doctor will get the person's medical history, which describes the person's problems and symptoms. A family history, which is a medical history of close relatives, is also helpful. The history plays a big part in pointing the doctor to CF because CF is a genetic disease.

Physical Examination

The doctor will do a full physical examination to look for CF ***signs*** or other problems.

Laboratory Tests

Laboratory tests are needed to confirm CF and to find out how CF has affected parts of the body. The ***sweat test*** is the standard test for CF, and a genetic test can diagnose CF as well. Details about CF diagnostic tests are found later in this chapter.

When Does CF Start?

CF starts at conception (before birth). However, CF symptoms may not show up for a while. The start of symptoms varies from person to person. For most people with CF the disease is diagnosed or symptoms begin to show up in the first 2 years of life.

About 10 percent to 15 percent of people with CF have symptoms at birth. They have a ***blocked intestine***, which is called ***meconium ileus***. When testing newborns, CF can usually be diagnosed even before symptoms show up.



People are born with CF, but some will not show any symptoms of the disease until later in their lives. Most children are diagnosed within the first 2 years of life. However, some people are not diagnosed until the teen years and even adulthood.

CF Symptoms

The type and severity of CF symptoms varies from person to person. Many other health problems have symptoms like those of CF. This can make CF hard to identify, or diagnose. CF may be mislabeled or undiagnosed for years.

Common CF symptoms:

- Salty-tasting skin
- Slow weight gain, even with good appetite
- Abnormal bowel movements (chronic ***diarrhea***; large, frequent, greasy, foul-smelling ***stools***)
- ***Wheezing***
- Cough and increased lung mucus
- ***Pneumonia***
- ***Nasal polyps*** (small fleshy growths in the nose)
- ***Clubbing*** (enlargement) of the fingertips and toes
- ***Rectal prolapse*** (the ***rectum*** sticks out through the ***anus***)

These symptoms are not seen only in people with CF. They may be seen in other illnesses. Someone with CF may not have all of these symptoms. When many of these symptoms are present, however, the doctor may want to do a test for CF. The standard test to diagnose CF is the sweat test.

Later chapters will cover how CF affects the body. They also cover more about CF symptoms, why they occur and how they are treated.



A sweat test is the standard way to diagnose CF. The test should be done at a Cystic Fibrosis Foundation–accredited care center. These care centers specialize in the diagnosis and treatment of CF.

Sweat Test

CF is usually diagnosed by a sweat test. The sweat test measures the amount of salt in a person's sweat. CF affects exocrine glands. Sweat glands are one kind of exocrine gland. The CF gene mutation changes how the body handles salt. More salt is lost in sweat. The amount of sweat is normal, but the salt in it is high. This is the basis for the test used to diagnose CF.

The sweat test is an easy, accurate way to diagnose CF. The sweat glands are made to produce sweat with a mild chemical called pilocarpine and a little electricity (***pilocarpine iontophoresis***). A gauze pad or piece of filter paper is put on the skin to collect the sweat. Then, the area is wrapped in plastic for 30 minutes. A small plastic strap that looks like a wristwatch also may be used to collect the sweat. It collects the sweat into a small tube. If the person has CF, the sweat will be high in salt (sodium and chloride). If the sweat test shows increased salt, the doctor will do a second sweat test to be sure.

The sweat test is easy, painless, accurate and inexpensive. But it must be done and analyzed correctly. Sweat tests must be done by trained, experienced people at a CF Foundation–accredited care center. Laboratories at CF Foundation–accredited care centers do many sweat tests each year, and have their sweat tests reviewed by a CF center director.

Other Facts about Sweat Tests

Sweat tests can only diagnose CF. They cannot predict the severity of CF. There is no connection between sweat salt level and how severe the disease is.

Sweat tests cannot identify CF carriers. Carriers do not have CF, so the salt in their sweat is normal.

What about Testing Relatives?

Because CF is genetic, ***siblings*** of a person with CF should be tested. Siblings should be tested whether or not they show symptoms of CF. The age when symptoms start varies from person to person. Finding CF early means that treatment can begin early. Early treatment can mean better health overall.

Other relatives (such as first cousins and half-siblings) should be sweat tested if they have any CF symptoms or if the family is concerned that they may have CF. This can be done with a sweat test. It may also be done with a gene test.

Gene Tests

About 90 percent of the time, gene tests can diagnose CF and tell which gene mutations are present. This can be done with a blood sample or cells brushed from the inside of the cheek (buccal smear). Half the people with CF have two copies of the F508del (sometimes written Delta F508 or ΔF508 mutation). You may hear this called homozygous for F508del or F508del/F508del. Most others with CF have one F508del mutation and one other CF gene mutation. A person who has one F508del and one of the hundreds of other CF gene mutations is heterozygous for F508del.

The genes are found by tests done in laboratories that specialize in genetic testing. There is some relation between the gene mutation and CF severity, but it is not simple to predict. CF varies even in people with the same gene mutations. This may be caused by other genetic and non-genetic factors. Knowing which CF gene mutations a person has does not give information about how healthy the person will be, how well they will do in life or how long they will live.

Recent research has shown that some treatments work for people with certain gene mutations. Therefore, knowing someone's specific CF gene mutations may be important in choosing therapies for that person.

Newborn Screening and Diagnosing Newborns with CF



Newborn Screening for CF

Newborn screening (NBS) is a nationwide program to identify babies who may have certain health conditions. CF screening tests newborns to identify babies who are at high risk of having the disease. If a baby has a positive NBS for CF, the baby will need to have a sweat test to see if he or she actually has CF.

By the end of 2009, all states included CF in their NBS tests. NBS is done so that the diagnosis of CF can be made early. Starting treatment early is important to keep the child as healthy as possible. Early treatment can lead to improved growth, keeping lungs healthy, fewer hospital stays and a longer life.

The NBS test is done during the first 2 to 3 days after birth, usually by the health care provider in the hospital. Most states check for a chemical made by the pancreas, an organ that helps with digestion. This chemical is

called ***immunoreactive trypsinogen***, or IRT. IRT is normally found in small amounts in the body. When the pancreas is stressed before a baby is born, more IRT is released into the baby's blood. IRT can be high if a baby is premature or had a stressful delivery, or for other reasons. IRT also tends to be high in people who have CF.

Some states use an IRT/IRT, or "IRT-only" method when they screen newborns for CF. If the first IRT is high, it is considered "positive," or abnormal. Babies with a high-level IRT from the blood collected in the hospital need to have a second IRT test at the doctor's office.

Most babies with a high IRT do not have CF. A positive or abnormal IRT result simply means the baby could have CF. If the second IRT test remains high, the parents are asked to bring the baby to a CF Foundation–accredited care center for a sweat test. The sweat test is used to diagnose, or to see, if the baby has CF.

Most states now use an IRT/***DNA*** method for NBS tests. If the IRT is high from the blood sample taken in the hospital, a second test is done on the same blood sample to check for some of the changes in the gene that causes CF. This is called a DNA test. When an IRT or IRT/DNA test is positive, the sweat test is used to see if the baby has CF.

Some babies that have a positive NBS test for CF do not have CF. This is called a ***false-positive***. Some babies with a negative NBS test for CF do have CF. This is called a ***false-negative***. Therefore, anyone, at any age, who has symptoms of CF should have a sweat test to see if they have CF.

Diagnosing Newborns with CF

When babies have a positive CF NBS test, the sweat test is used to make the diagnosis, or to see if the baby actually has CF. The sweat test should be done at a CF Foundation–accredited care center. The sweat test can often be done on an infant as young as 2 weeks old. Sometimes babies do not make enough sweat to get an accurate sweat test. In those cases, the sweat test has to be done when the baby is older and enough sweat can be collected for the test.

Can CF Be Diagnosed Before Birth?

Some genetic conditions can be found before birth, or prenatally. CF can sometimes be diagnosed before birth. This can be done by ***amniocentesis*** or ***chorionic villus sampling*** (CVS). In amniocentesis, a small amount of fluid around the fetus is tested. In CVS, a small piece of ***placenta*** is tested. The chromosomes in cells from an amniocentesis or CVS are studied for known CF gene mutations. Also, CF can be diagnosed by looking at proteins in ***amniotic fluid***.

Couples who are carriers of the CF gene who do not want a child with CF can consider ***in vitro fertilization.*** The embryo can be tested for CF before it is implanted in the mother.

These prenatal tests are not 100 percent accurate. Standard prenatal genetic screening tests look for only the most common CF gene mutations. There are more than 1,500 CF gene mutations. The parent or parents could carry a gene mutation that was not included in the prenatal testing. Because of this, infants should still have a CF NBS test done.

Remember

False-positive and false-negative results do occur with CF NBS. All infants with a positive NBS test should have a sweat test.

Although CF symptoms may start at any age, anyone with symptoms of CF should have a sweat test done for diagnosis, even if they have had a negative NBS.

In some cases, a genetic test to diagnose CF can be done instead of a sweat test. A person has to have two abnormal CF genes or CF mutations to be diagnosed with CF.

Because CF is inherited from the parents, siblings of a person with CF should be tested. Because both parents inherited CF gene mutations from their parents, siblings of the parents may also carry the gene. First cousins of the person with CF should be tested if they have CF symptoms.

Early diagnosis is important so that medical and nutritional treatments can be started as soon as possible. Early treatments lead to improved long-term health.

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **A person who has symptoms of CF should have a sweat test to see if he or she has CF.**
   a. True
   b. False

2. **Which of these symptoms are seen only in CF and not in any other disease?**
   a. Cough
   b. Wheezing
   c. Reoccurring lung infections
   d. Difficulty gaining weight
   e. All of the above
   f. None of the above

3. **Which kind of test is used to diagnose CF?**
   a. A test to check mental skills
   b. A test to check muscle coordination
   c. A test to check for too much salt in the sweat
   d. A test to check for congestion in the nose and lungs

4. **Which of these are true about the sweat test for CF?**
   a. The test is simple, painless and reliable.
   b. The test cannot identify carriers of the CF gene.
   c. The test can tell if the person has a mild or severe case of CF.
   d. The test should be done by a medical lab that does many of these tests every year, such as a CF Foundation–accredited care center.

5. **When a person is found to have CF, who else also should be tested?**
   a. The parents of the person
   b. Brothers and sisters of the person
   c. Anyone who has had close contact with the person

6. **Which of the following tests may be used to diagnose CF?**
   a. Sweat test
   b. Gene test using a blood sample
   c. Gene test using cells from the inside the cheek
   d. All of the above

Answers

To learn more, turn to the page number shown after the answer.

1. A
It is **true** that a person who has symptoms of CF should have a sweat test to see if he or she has CF. Page 24.

2. F
None of the above symptoms are seen only in CF. These symptoms are seen in other diseases as well as CF. Page 24.

3. C
A test to check for too much salt in the sweat is used to diagnose CF. Mental skills and muscle coordination are not affected in CF. Congestion in the nose and lungs occurs in many people who do not have CF. Page 25.

4. A, B, D
It is true that **the test is simple, painless and reliable; it cannot identify carriers of the CF gene** and **it should be done by a medical lab that does many of these tests every year, such as a CF Foundation–accredited care center**. The sweat test can only tell if a person has CF. The amount of salt in the sweat does not tell you if the person has a mild or severe case of CF.
Page 25.

5. B
When a person is diagnosed with CF, the **brothers and sisters of the person** also should be tested. The symptoms of CF begin at different ages for different people. Brothers and sisters could have CF and not know it. Because CF is inherited and not contagious, there is no reason to test anyone simply because they have had close contact with a person with CF. Page 23.

6. D
All of the above tests can diagnosis CF. The sweat test is used most often. A positive sweat test is always repeated to confirm the diagnosis. Gene testing also can be done or may be used if the sweat result is unclear. Gene testing can be done on either blood or cheek cells. Pages 25-26.

Notes and Questions

PART II

How CF Affects the Body



Sweat Glands

Cystic fibrosis (CF) affects the exocrine glands. It causes these glands to make abnormal secretions. The secretions made by most exocrine glands in a person with CF are thick and sticky. The thick and sticky secretions block organ passageways. How CF affects the sweat glands is different than how CF affects other exocrine glands.

How CF Affects the Sweat Glands

Sweat glands are exocrine glands, but the sweat in CF is not thick and sticky. The sweat in CF is normal except for being too salty. This is because of how the sweat glands work in someone with CF. There is two to five times more salt in the sweat of a person with CF. This high salt level is the reason a person is tested for CF with the sweat test.

The Symptoms

The high amount of salt in a person's sweat is a classic sign of CF. The family is often the first to notice this sign. When they kiss the child, the child's skin may taste salty. People with CF lose more salt than normal in their sweat. For people with CF, it is rarely a problem because there is usually enough salt in food to replace the salt they lose in their sweat. But, when they sweat more, they lose more salt, and that can be a problem.

People sweat more:

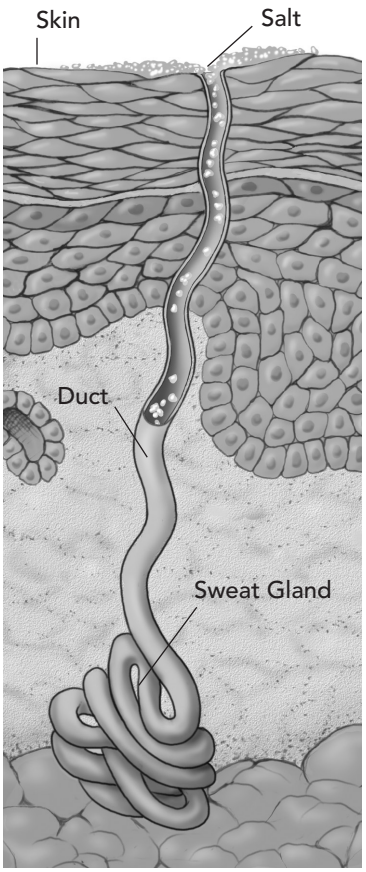
- In hot weather
- When they exercise
- With a fever

The symptoms of losing too much salt are:

- Tiredness
- Weakness
- Vomiting
- Fever
- Muscle cramps
- Stomachache

Losing too much salt can lead to:

- *Dehydration*
- *Heat stroke*



Sweat produced in a person with CF has a very high level of salt.

Treatment

Losing too much salt is an easy problem to treat. People with CF should eat salty foods and add salt to other foods. People with CF should use salt freely. They should have their own salt shaker. Having unlimited salt helps avoid problems.

When sweating more (in hot weather, during exercise or with a fever), someone with CF should drink more and eat more salty food. If symptoms of salt loss are present, the doctor may give extra salt.

People with CF may need to avoid some times of heavy sweating but should not limit their activity. No one should have too much sun or heat. It is important to use common sense.

Remember

Everyone with CF loses more salt in their sweat than normal. When sweating more (in hot weather, during exercise or with a fever), people with CF should drink more and take more salt. Learn the symptoms of salt loss in this chapter. If you see these symptoms, call the doctor. Very few people with CF have to watch how much salt they eat.

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **CF affects sweat glands by:**
   a. Increasing the amount a person sweats
   b. Increasing the amount of salt in sweat
   c. Making the sweat thick and sticky

2. **Which of these is a symptom of losing too much salt?**
   a. Fever
   b. Muscle cramps
   c. Tiredness and weakness
   d. Stomachache and vomiting
   e. Heat stroke and dehydration
   f. All of the above

3. **Which statement about CF and hot weather is not true?**
   a. A person with CF should eat salty snacks.
   b. People with CF should never play in hot weather.
   c. Most people with CF do a good job of adjusting their salt intake.
   d. Extra fluids and salt may be needed when a person with CF is sweating more.

Answers

To learn more, turn to the page number shown after the answer.

1. B
CF affects the sweat glands by **increasing the amount of salt in sweat.** The amount of sweat in CF is normal and it is watery like the sweat of someone without CF. Page 35.

2. F
All of the above are symptoms of losing too much salt. Page 35.

3. B
It is **not true** that **people with CF should never play in hot weather**. People with CF do well in hot weather as long as they drink more, eat salty snacks and do not overdo it. Page 36.

Notes and Questions

Respiratory System

Breathing problems (also called respiratory or ***pulmonary*** problems, which affect the lungs and the airways) are the most serious problems associated with cystic fibrosis (CF). When breathing problems are first noticed is different for each person with CF. How severe these problems are will also differ for each person. For most people with CF, lung disease affects how well they do and how long they live. Many treatments are available to keep the lungs as healthy as possible.

How the Respiratory System Works

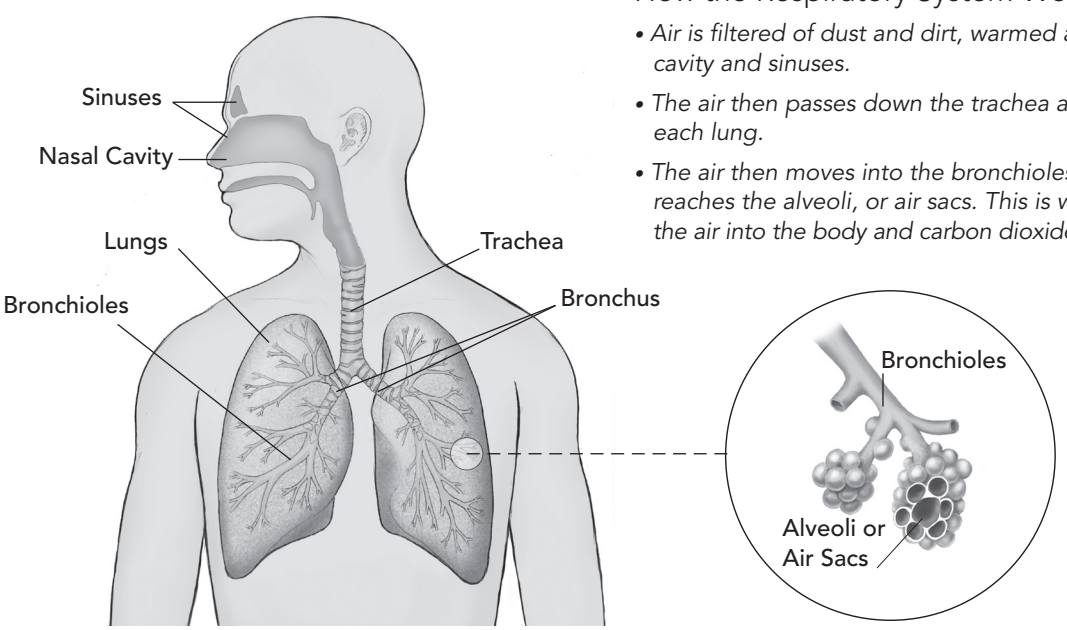
The ***respiratory system*** has two main parts:

1. ***Upper respiratory tract*** — the nose and ***sinuses***

   The upper respiratory tract filters out particles such as dust and germs in the air we breathe. It also warms and moistens the air.

2. ***Lower respiratory tract*** — the airways and lungs

   The lower respiratory tract starts at the ***trachea*** (the main windpipe).



How the Respiratory System Works

- *Air is filtered of dust and dirt, warmed and moistened in the nasal cavity and sinuses.*
- *The air then passes down the trachea and into the bronchus of each lung.*
- *The air then moves into the bronchioles or smaller airways until it reaches the alveoli, or air sacs. This is where oxygen is taken from the air into the body and carbon dioxide is removed from the body.*

The trachea branches into one ***bronchus*** (breathing tube or bronchial tube) for each lung. The bronchus then branches into bronchial tubes that spread through the lungs like tree branches. The smaller branches are called ***bronchioles***. The smallest branches end in tiny air sacs called ***alveoli***. These air sacs inflate and deflate like tiny balloons with each breath. It is here that gas exchange occurs. Gas exchange is getting oxygen into the body and waste gases out of the body.

The upper airways filter out most of the particles in the air we breathe. But some do get through and into the lower respiratory tract. If allowed to stay, they can block the tiny air tubes. This can allow germs to grow and cause lung infections.

Exocrine glands in the airways help remove things such as dust and germs that get into the lower respiratory tract. These glands make thin, slippery mucus that lines the inside of the airways, trapping particles and germs. Under the mucus, the airways have cells with special cleaning tools called ***cilia***. The cilia are like small hairs. They move together to push the mucus to the throat. The particles and germs we breathe in are trapped in the mucus and then carried to the throat where they can be coughed out or swallowed.

The main purpose of the respiratory system is gas exchange. Gas exchange has two parts:

1. Oxygen exchange — the respiratory system takes oxygen from the air we breathe and transfers it to the blood. Oxygen is then carried to all body parts.
2. Waste gas exchange — the respiratory system transfers the waste gas (carbon dioxide) from the blood to the air we breathe out.

The Problem

CF affects both the upper and lower respiratory tracts. The most important effects of CF are to the lower respiratory tract.

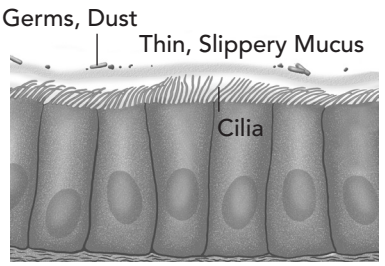
Lower Respiratory Tract

In CF, the airway mucus is too thick and sticky. It traps particles, but the mucus is so thick and sticky that the cilia cannot easily move the mucus to the throat. So, instead of cleaning the airways, the mucus clogs them. When mucus blocks an airway, it is called a mucous plug.

Mucous plugs keep air from getting into or out of some alveoli (air sacs). Oxygen cannot get into the blood and carbon dioxide cannot get out. The thick, sticky mucus in the small airways:

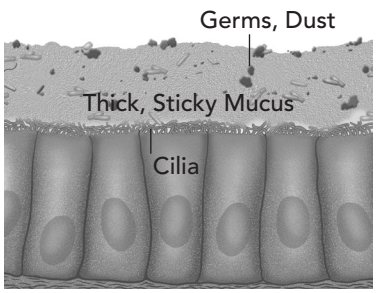
- Gets in the way of air getting into and out of alveoli, and
- Makes it hard to remove particles and germs from the airways.

Healthy Airways



Healthy airways have a thin layer of mucus, which can easily be moved to the throat by the cilia and then coughed out or swallowed.

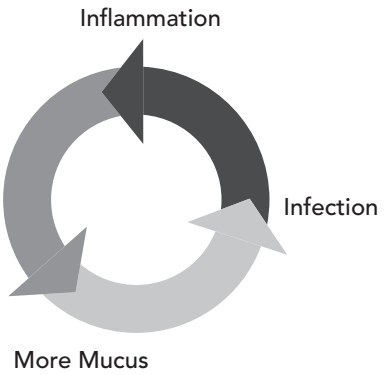
The Problem



In CF, the airway mucus is thick and sticky, which stops the cilia from moving the mucus to the throat to be coughed out or swallowed.

If not removed, mucous plugs can lead to lung infections and lung damage. Lung infections cause problems for people with CF because:

- Infections cause inflammation, which can damage the cilia and airways, making them even less able to clear mucus, particles and germs from the lungs.
- Infections often cause more mucus to be made. This can lead to more mucous plugs and worse infections.



The infection–inflammation cycle causes lung damage in CF.

This infection–inflammation cycle causes lung damage. Mucus plugs airways and germs grow, causing infection. The lungs react by making more mucus to clean the airways, but the mucus plugs more airways. The infection grows more and the cycle repeats.

People with CF are at risk for certain lung infections. There are many ***bacteria*** or germs that cause CF lung infections.

Two are:

- ***Staphylococcus aureus*** (or simply staph, pronounced "staff")
- ***Pseudomonas aeruginosa*** (or simply *Pseudomonas*, pronounced "soo-doe-mow-nas")

Treatment of CF in the lower airways focuses on clearing the airways and treating the bacteria in the lungs. (See the Treatment section later in this chapter on page 46.)

Upper Respiratory Tract

CF also affects the upper respiratory tract — the nose and sinuses. Although CF does not often cause big problems there, two conditions may occur:

Sinusitis

Sinusitis can occur in CF. Sinus inflammation is caused by thick, sticky mucus blocking the sinuses. This blockage may lead to sinus infections. Sinusitis may be treated with ***antihistamines*** and decongestants to open up the sinuses, and with ***antibiotics*** to control infections. Sinus irrigation (flushing) or other treatment may be needed. Sometimes, surgery may be needed to open up and drain the sinuses.

Nasal Polyps

Nasal polyps are small, fleshy growths in the nose. These are less common than sinusitis. We do not know what causes polyps. They may be caused by problems with mucous glands in the nose. Nasal polyps do not always need treatment. If they block the nose or cause problems, then medicines or surgery may be needed.

Symptoms

Most of the respiratory symptoms of CF are caused by thick mucus blocking airways. When respiratory symptoms of CF are first noticed, and how severe they are, is different for each person. The symptoms increase over time as CF lung disease gets worse. From time to time, the lungs of a person with CF get worse. This is called a pulmonary ***exacerbation*** or flare-up. Lung infections cause exacerbations. A person with CF will need extra treatments during an exacerbation.

It is important to know what symptoms are normal for a person with CF, and then to notice when the symptoms are worse. If CF respiratory symptoms become worse, the person with CF usually needs treatment. New or worsened symptoms should be reported to the CF care provider early, so that treatment can begin early.

Some symptoms are:

- Coughing
- Coughing up mucus (***sputum, phlegm***)
- Wheezing
- Repeated lung infections (***bronchitis*** or pneumonia)
- Shortness of breath or labored breathing (cannot "catch" your breath)
- Difficulty exercising, getting tired or needing to be less active

Tests

Your CF care team learns a lot by hearing your report of symptoms and how the person with CF is feeling and acting. The physical exam gives them more information. Your CF doctor may want to do some tests to learn still more. Tests are done on a routine basis. They are also done when a person may be having more symptoms. There are four kinds of tests that help your CF doctor know how your lungs and airways are doing.

Chest X-Ray

Chest X-rays let doctors look inside the lungs. X-rays show how CF is affecting the lungs. A chest X-ray will be done about every 1 to 2 years, and when a person is having more breathing problems.

Pulmonary Function Tests

Pulmonary function tests (PFTs) or breathing tests measure lung function, or how well the lungs are working. PFTs give information about airways that are blocked by mucus. They measure how fast and how much air can get in and out of the lungs. PFTs help the doctor see lung changes over time and help guide treatment. PFTs can also measure how the airways respond to inhaled medications.

Routine breathing tests may not start until a child is 5 or 6 years old, though some children may be able to do PFTs at an earlier age. It is hard to do these tests on infants and young children. However, some CF care centers may have the special equipment to be able to do these tests in younger children and infants.



Pulmonary function tests help CF doctors to see how well the lungs are working or if there are problems.

Sputum Cultures (Phlegm or Mucus Cultures)

Cultures check sputum, or mucus from the lungs, for germs that cause lung infections. Some mucus is put in a dish with a ***culture medium*** that helps bacteria such as staph and *Pseudomonas* grow. Later, the dish is checked for these and other bacteria that cause lung infections. Tests can then be done to see which antibiotics will work to treat that bacteria and which ones the bacteria would be able to resist. Antibiotics are medicines that fight the bacteria.

When a person is older, he or she can usually cough up some sputum. Infants and younger children cannot "expectorate," or cough up mucus. If a person cannot cough up some sputum or phlegm, an oropharyngeal (OP) swab or culture is done. Some centers call this a throat culture, or a "gagged" ***sputum culture*** and some may call it a "deep throat culture." To get a good sample, the throat culture swab should make the person gag so some sputum comes up in the throat from the lungs.

Blood Tests

Blood tests are done to help check the health of a person with CF. A small sample of blood is usually taken with a syringe and sent to a laboratory for testing.

Blood tests can be done to check:

- Nutrition status
- Vitamin levels
- For liver disease
- For ***diabetes***
- For infections
- The body's response to infection
- To make sure medicines are at the right level in the body
- Possible side effects of medicine

Treatment

CF lung disease begins early and often happens in the first few months of life. Lung disease begins even before any symptoms are noticed. Many treatments are started even before symptoms are noticed. These chronic or long-term treatments are aimed at keeping the lungs as healthy as possible. The goals of treatment are to clear blocked airways and to treat or prevent infections.

Because CF affects people differently, treatment is specific to each person. Your doctor and CF care team will work with you to develop the best treatment plan.

When you perform the daily, chronic treatments, you are working to prevent exacerbations or flare-ups and the damage they can cause to the lungs and airways. If an exacerbation happens, treating it as early as possible also helps keep the lungs healthier. Know what your or your child's normal symptoms are, and contact your CF care center if you notice an increase or change in these symptoms.

Chronic Treatments

Chronic treatments are essential to keep the lungs as healthy as possible. Some chronic treatments will be described here. Talk to your CF care provider and team to see what the best chronic treatments are for you or your child.

Airway Clearance Techniques

Airway clearance techniques (ACTs) are treatments that help people with CF stay healthy and breathe easier. ACTs are recommended for all people with CF. ACTs loosen thick, sticky lung mucus so it can be cleared by coughing or ***huffing***. Clearing the airways reduces lung infections and improves lung function.

There are many ACTs. Most are easy to do. For infants and toddlers, ACTs must be done by the parent. Older kids and adults can do their own ACTs. ACTs are often used with other

treatments, such as inhaled ***bronchodilators*** and antibiotics. Bronchodilators should be taken **before** or with ACTs to open airways. Inhaled antibiotics should be taken **after** ACTs to treat opened airways. Your CF care team will help you choose the best ACT and other treatments. Each year, review and update your routine with your CF care team. Different types of ACTs are described next.

Huff Coughing

Huff coughing is the most basic ACT. Huff coughs are the most effective coughs. Huff coughing involves taking a breath in and actively ***exhaling***. It is like "huffing" onto a mirror or window to steam it up. It is not as forceful as a cough but can work better and be less tiring. Good strong huff coughs help get rid of mucus.

People with CF should cough. Coughing is helpful and should be encouraged. People with CF should never try to suppress coughs or keep themselves from coughing. People with CF should not use cough suppressants. Coughing is a healthy, natural way for the lungs to get rid of mucus. Always cough into a tissue, throw it away in a trashcan and then clean your hands.

Chest Physical Therapy or Postural Drainage and Percussion

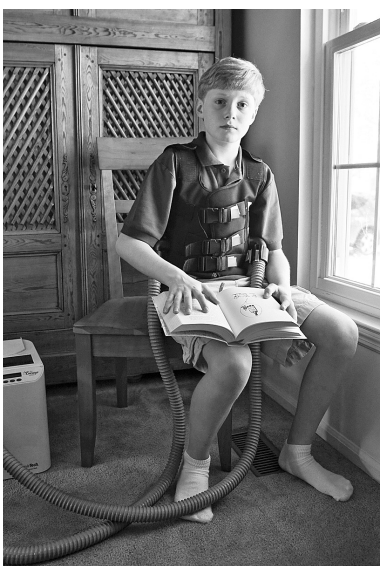
Chest physical therapy (CPT** or **Chest PT) or ***postural drainage*** and ***percussion*** (also called PD & P) is an ACT. With postural drainage, the person gets in several different positions (postures) that help drain mucus from different lung parts. Gravity pulls mucus from small to large airways where it can be coughed out. With chest percussion, the chest is clapped and vibrated to dislodge and move mucus. This is done in a variety positions to drain all lung parts. To learn more, go to Appendix C , Page 187, and read "An Introduction to Postural Drainage and Percussion."

High-Frequency Chest Wall Oscillation

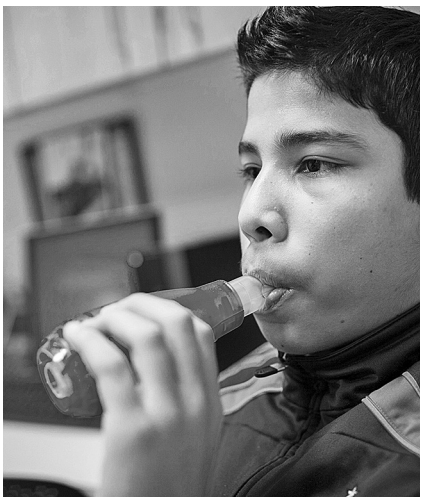
High-frequency chest wall oscillation also is called a "vest" or "oscillator." An inflatable vest is attached to a machine that vibrates it at high frequency. The vest vibrates the chest to loosen and thin mucus. Every 5 minutes the person stops the machine and huff coughs.

Positive Expiratory Pressure Therapy

Positive expiratory pressure (PEP) therapy gets air into the lungs and behind the mucus in the (collateral) airways. PEP holds airways open, keeping them from closing. A PEP system includes a mask or mouthpiece attached to a resistor set by your CF care team. The person breathes in normally and breathes out a little harder against the resistance.



Many people with CF use a vest that helps them cough the thick, sticky mucus out of their lungs.



Oscillating PEP

Oscillating Positive Expiratory Pressure

Oscillating positive expiratory pressure (oscillating PEP) is an ACT where the person blows all the way out many times through a device. Types of oscillating PEP devices include the Flutter,™ Acapella,™ Cornet™ and ***intrapulmonary percussive ventilation (IPV)***. Breathing with these devices vibrates the air in large and small airways. This vibration dislodges and moves mucus. After blowing through the device many times, the person huff coughs. This cycle is repeated many times.

Active Cycle of Breathing Technique

Active cycle of breathing technique (ACBT) involves a set of breathing techniques. It can be changed to meet each person's needs. It gets air behind mucus and clears mucus. ACBT includes:

- Breathing control — normal, gentle breathing with the lower chest while relaxing the upper chest and shoulders.
- Chest expansion exercises — deep breaths in. Some use a 3-second hold of breath to get more air behind the mucus. This may be done with chest clapping or vibrating, followed by breathing control.
- Forced expiration technique — huffs of varied lengths with breathing control.

Autogenic Drainage

Autogenic drainage (AD) means "self-drainage." It uses different speeds of breathing to move mucus. This moves mucus from small to large airways. AD has three parts:

1. Dislodging mucus
2. Collecting mucus
3. Clearing mucus

The person inhales to different levels and then adjusts how he or she breathes out to change how fast the air flows out of the lungs and moves mucus. At first, AD takes hard work and practice. It is best for people who are older than 8 years of age.

The best ACT method is the one that works best for the person with CF. Talk to your CF care provider, respiratory therapist or physical therapist. They can help you choose what is best for you and learn to do the technique accurately. If you have questions, please contact your local CF care center.

The CF Foundation also has information about airway clearance on its website at **www.cff.org** or you can call **1-800-FIGHT CF.**

Exercise

Everybody needs exercise. People of all ages with CF should exercise and be active. Most people with CF can do any exercise and play sports and games (except scuba diving) that they want. Exercise can:

- Loosen mucus in the lungs so it can be coughed up easier
- Cause coughing, which helps clear the lungs
- Strengthen breathing muscles
- Strengthen the heart

Good Nutrition

Good nutrition is very important for people with CF of all ages. There is a connection between good nutrition and good lung function. A balanced high-calorie diet with fat and protein gives the body what it needs to grow normally.

Normal gains in weight and height help:

- Build strong lungs
- Keep lungs as healthy as possible
- Build and keep a strong immune system to fight infections

Medications

Bronchodilators

Bronchodilators open the airways to help clear mucus and make breathing easier. Bronchodilators are given by breathing in a medication. Albuterol is a bronchodilator.

Mucolytics

Mucolytics are medications that thin mucus, making it easier to cough out. Pulmozyme® (DNase) is a mucus-thinning medication that is taken by inhaling, usually once a day.

Hypertonic Saline

Hypertonic ***saline*** is extra-salty water that is sterile, so there are no germs in it. CF airways do not have enough salt and water in them. Hypertonic saline is inhaled in the lungs as a mist, which helps to clear the thick mucus from the lungs.

Azithromycin

Azithromycin, also known as Zithromax,® is a commonly used antibiotic. It is a pill that is is taken by mouth 3 times a week. Azithromycin has been shown to improve lung function when taken regularly. Many people without CF have taken this drug to treat pneumonia, sore throats or ear infections.

Many CF medications are aerosol treatments and inhaled into the lungs.



Ibuprofen

Ibuprofen is an ***anti-inflammatory*** and non-steroidal agent (does not affect hormonal balance). It is taken as a pill. Ibuprofen has been used in some people with CF and has been shown to slow the rate of lung disease.

Ibuprofen is not right for everyone with CF. There are several factors to consider before taking this drug long term. When ibuprofen is taken for CF, the doctor needs to prescribe the right amount. The doses needed are quite high and are based on a blood test.

Do **not** start taking ibuprofen from the drug store for CF without talking to your CF doctor. Too low a dose means that you or your child will miss the benefit and still risk side effects. There is an even greater risk if the dose is too high. Talk to your CF doctor to find out if ibuprofen is right for you or your child.

Ivacaftor

In CF, the gene mutation changes the CFTR protein so it doesn't work properly. Kalydeco™ (ivacaftor) helps the CFTR protein work in people with at least one mutation called G551D. Kalydeco is a pill taken by mouth twice a day for the treatment of CF in people with this mutation ages 6 years and older. It helps improve lung function, lower sweat chloride levels and increase weight gain. Research is being done to find out what other CF mutations Kalydeco might help work.

More research is being done to find more medicines that will help the CFTR protein work for people with CF with other mutations, like those with at least one F508del (sometimes called Delta F508). You can learn more about this research and different mutations of CF on the CF Foundation's website at **www.cff.org**.

Inhaled Antibiotics

Inhaled antibiotics are used to fight or control infections. The CF care provider uses the results of the sputum culture to see if an inhaled antibiotic should be taken.

Inhaled antibiotics go right to where they are needed — the lungs. The antibiotics especially made for people to inhale are tobramycin ***inhalation*** solution (such as TOBI®), Cayston® (aztreonam for inhalation solution) and TOBI® Podhaler™ (tobramycin inhalation powder). These antibiotics are used to improve respiratory symptoms in people with CF who have *Pseudomonas aeruginosa*.

How to Give Inhaled Medications

Inhaled medicines are given by ***aerosol, dry powder inhaler (DPI)*** or ***metered dose inhaler (MDI)***.

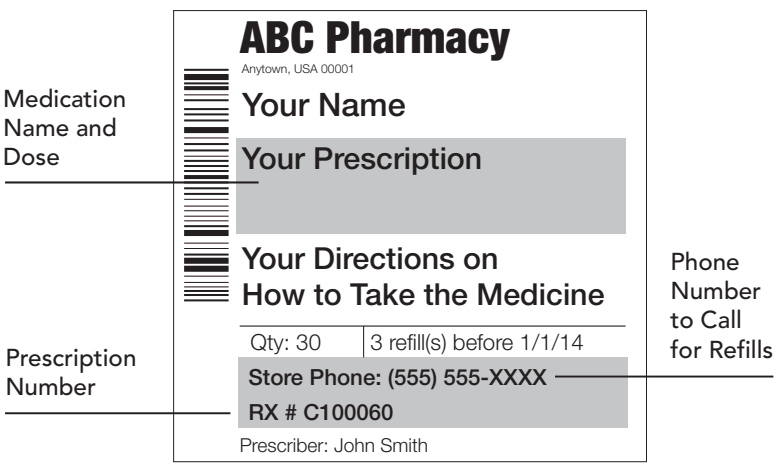
Aerosols are mist treatments made from liquid medicines. The drug is put in a cup (called a ***nebulizer***) and attached to a small ***air compressor***. Some of these medicines are mixed with saline (salt water) in the nebulizer. The compressor blows air through the nebulizer cup making a mist. The mist is inhaled through a mouthpiece or mask for several minutes. Some inhaled medicines may be used with ACTs to help clear the mucus from the lungs.

Some inhaled medicines are given through a DPI. This medicine is a powder that is inhaled using a small device. Medicine given by an MDI use a small device also. An MDI gives one dose of medicine at a time as a spray. When using an MDI, always use a spacer or holding chamber.

There are several different types of nebulizer and compressor devices. Certain aerosol medications require that specific devices are used. Similarly, different DPI and MDI devices use different techniques in order to properly get the medicine in the lungs. Your CF care team can teach you how to use these devices correctly.

How to Refill Prescribed Medications

Many people with CF use medications every day. This means that you will need to know when and how to get your medicine refilled. You should order your refills about 8 to 10 days before you run out of medicine. This allows your pharmacy to get the medicine or, if you are using a mail-order pharmacy, for it to be mailed to you. The information about how to refill your medicine is on the label of the medicine bottle or box.



Steps to refill prescriptions:

- Order refills 8 to 10 days before you run out.
- Have the label handy when you are ordering refills.
- On the label, find either the telephone number or Web address you can use to order the refill.
- Give the prescription number when you order.
- If you are using a mail-order pharmacy, make sure the pharmacy has your right mailing address.

Treatment for Exacerbations

Sometimes CF symptoms may get worse or you may notice new symptoms. This could be a pulmonary exacerbation or flare up, which may need extra treatment. Call your CF care center if you think you or your child are having a pulmonary exacerbation so that treatment can start right away and lung damage can be less.

Symptoms of a pulmonary exacerbation may include any of the following:

- More coughing and/or wheezing
- Chest congestion
- More mucus (sputum, phlegm)
- Change in sputum color (to dark yellow or green)
- Tiredness, less energy
- Being less able to exercise
- Weight loss and/or poor appetite
- Sometimes blood-streaked mucus (always let your CF care provider know about blood in the mucus)
- Sometimes fever

Treatment for a pulmonary exacerbation includes antibiotics, more airway clearance and good nutrition. Antibiotics may be given by mouth, also called oral, or ***intravenous***, also called ***IV***. Oral antibiotics are liquids, tablets or capsules that are swallowed. Oral antibiotics are used to treat mild pulmonary exacerbations. They work against many bacteria, but do not always work against *Pseudomonas*.

IV antibiotics are often necessary for CF pulmonary exacerbations. IV antibiotics are solutions with antibiotics that are put into the blood through a small tube called an IV catheter. They are needed if oral antibiotics do not work, or if the infection is caused by bacteria that are only sensitive to an IV antibiotic. People with pulmonary exacerbations who need IV antibiotics are usually admitted to the hospital to receive the care they need. Sometimes, if they are able to get the extra airway clearance and nutrition at home, they can complete the IV antibiotics at home.

Possible Respiratory Complications of CF

There are other possible respiratory complications of CF, which are discussed here.

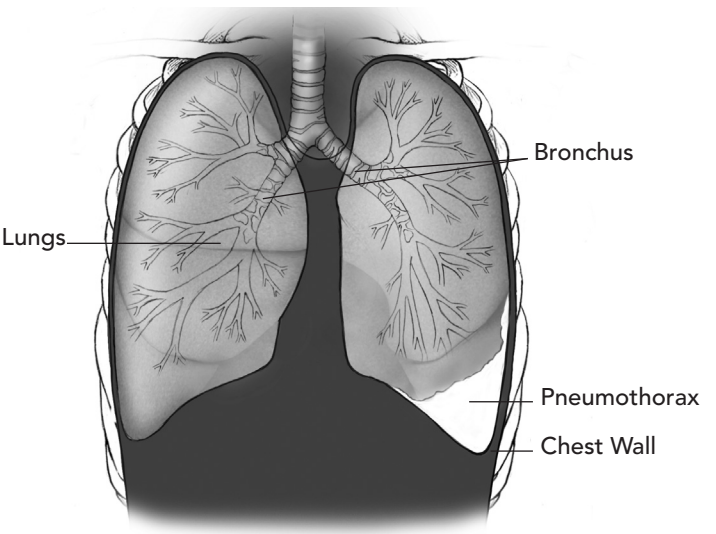
Bronchiectasis

In most people with CF, the airways get inflamed and damaged. This causes the airways to stretch and get floppy. This is called ***bronchiectasis***. It makes it harder to clear the airways of mucus and causes a change in breathing. Airway clearance techniques with huffing and coughing can help clear floppy airways of mucus.

Pneumothorax

Pneumothorax is a break in lung ***tissue*** or an airway that lets air escape from the lung. This air gets trapped between the lung and the chest wall. As more air escapes, it may cause the lung to collapse, making it harder to breathe. Call the CF doctor if there is sudden shortness of breath or chest pain!

Pneumothorax occurs in less than 4 percent of people with CF. With a mild pneumothorax, the air may re-enter the lung over time and you will not need medical help, or only need oxygen. A few people that have a pneumothorax may need more medical help to remove the trapped air so the lung can expand. Some people may need surgery to keep a pneumothorax from happening again.



A pneumothorax is air trapped between the lung and the chest wall.

Clubbing

Clubbing is when the tips of the fingers and toes get bigger and rounder. This is common in CF. The cause is not known, but it seems to occur with lung disease. Clubbing can change over time. It is not a sign of lung disease severity. Clubbing also occurs in people with heart disease and other types of lung disease.

Hemoptysis

Hemoptysis means to cough up blood. In CF, it may look like small blood streaks in the sputum. This may be a sign of an exacerbation that needs treatment. It is caused by bleeding from the lining of an airway. This lining may become inflamed and easily damaged. The thick, sticky mucus can then scrape or tear the lining, and cause bleeding.

Less often, hemoptysis may occur when an artery breaks and bleeds into an airway. This is more serious and may need immediate medical treatment or surgery. If the sputum has blood, call your CF care center.

Cor Pulmonale

In some people, after years of living with CF, a lot of lung damage may cause the right side of the heart to grow thicker and larger. This is called ***cor pulmonale***. The lung damage limits gas exchange, so less oxygen gets into the blood. To get the oxygen the body needs, the heart works harder to pump more blood through the lungs.

Also, lung damage may increase the ***blood pressure*** of the arteries going from the heart to the lungs. This makes the heart work harder to pump blood to the lungs. The strain on the heart makes the right side of the heart grow larger. Then the heart may tire and not pump as well. This may cause swelling, especially of the feet and ankles.

Cor pulmonale is treated by improving lung function and health. Extra oxygen and ***diuretics*** may help. Diuretics are medicines that get rid of extra fluid in the body.

Lung Transplantation

When someone with CF develops severe lung disease, the CF care team may discuss the option of lung transplantation with the person. The CF doctor can refer the person to a lung transplant center for evaluation. The transplant center evaluates the person's health to determine if a lung transplant is right for that person, and if it is the right time for it to be done.

Tests examine how well the lungs, heart and kidneys function, and the types of bacteria in the lungs. Because of the serious health care implications of transplantation, the person's psychological well-being is also evaluated. The transplant center will evaluate the person's social support system including family, friends and professional support. Most parts of the evaluation are standard, but each center can have some specific requirements. The decision whether to accept a person for a transplant is specific to that center.

As many as 90 percent of people with CF are alive 1 year after transplantation, and more than half are alive after 5 years. Following surgery, a person may be discharged from the hospital in a few days or a few months, depending on the person's health and complications. The average time in the hospital after transplant is 2 weeks.

The immune system protects the body from foreign material, which is anything not belonging in the body, such as germs or transplanted organs. Therefore, after the transplant, the immune system naturally reacts against — or rejects — the new organs. Drugs to stop the immune system from rejecting the organ, called immunosuppressive drugs, must be taken daily for life.

These drugs may cause side effects such as diabetes, kidney problems, cancer and ***osteoporosis*** (i.e., thinning of the bones). Research on immunosuppressive drugs shows promise in helping people live longer with lung transplants.

Transplanted lungs come from people who do not have CF, so the new lungs do not have CF. However, after the transplant, the recipient still has CF in the sinuses, pancreas, intestines, sweat glands and reproductive tract. The new lungs do not "get" CF, but immunosuppressive drugs may decrease the body's ability to fight germs that cause respiratory tract infections and other types of infections.

Lung transplantation was first done in the United States in the early 1980s. Over time, survival after a lung transplant has improved. The CF Foundation supports research on CF and transplantation and policies that help people with CF get lung transplants. It is hoped that the CF Foundation's efforts to develop new drugs for CF will reduce the need for lung transplants.

Making the decision to have a lung transplant is a very difficult and personal one. People with CF should talk to their CF care team members to get more information about lung transplant. Visit the CF Foundation website (**www.cff.org**) for more information.

Staying Healthy and Avoiding Germs

It is always better to prevent a health problem than to try to fix one after it has started. For instance, ***vaccinations (immunizations)*** are vital for prevention. Everyone should get the recommended vaccinations, **and** a yearly flu shot. Avoiding germs as much as possible is always best. Washing hands or hand hygiene should be done often. It is very important for people with CF to avoid exposure to tobacco smoke.

Read more about "Staying Healthy: What You Can Do" in Chapter 8, and "Staying Healthy: Avoiding Germs" in Chapter 9.

Remember

The effects of CF on the respiratory system require a variety of daily treatments including ACTs and inhaled medicines. Good nutrition and exercise are also important for respiratory health. Staying on track with the daily schedule of treatments — and talking to the CF health care team about any changes in symptoms — will help a person with CF keep his or her airways as strong and healthy as possible.

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **Which of these statements is true about lung disease in CF?**
   a. CF affects the lungs.
   b. Breathing problems are first noticed at different times for each person.
   c. Many treatments are available to keep the lungs as healthy as possible.

2. **The function of mucus in the lower airways is to:**
   a. Moisten the air we breathe
   b. Seal leaks that may occur in the smallest air passages
   c. Plug up bad or damaged airways and direct air to only the healthy parts of the lung
   d. Trap particles and germs so they can be carried toward the throat and coughed up or swallowed

3. **Which of these statements about the mucous plugs in CF is true:**
   a. Mucous plugs are uncommon in people with CF.
   b. Mucous plugs block the flow of air in the lower airways.
   c. If not removed, mucous plugs can lead to lung infections.
   d. Mucous plugs make it hard to remove particles and germs from the airways.

4. **Which of these symptoms occur when there is a pulmonary exacerbation?**
   a. Tiredness
   b. Weight loss and/or poor appetite
   c. More coughing and/or wheezing
   d. More mucus (sputum/phlegm)
   e. Any of the above.

5. **The purpose of airway clearance techniques in CF is to:**
   a. Clear the airways
   b. Prevent spread of CF to others
   c. Loosen the mucus so it can be coughed out of the lungs
   d. Wash out the lungs and cure CF

6. Which of these statements about exercise is true for people with CF?
   a. Exercise helps to loosen mucus in the lungs.
   b. Exercise helps strengthen breathing muscles.
   c. Exercise may cause coughing and help clear the airways.
   d. Exercise should be avoided because it may make the lung disease worse.
7. In CF, antibiotics are used to:
   a. Treat chronic lung infections, or control infections
   b. Treat acute pulmonary exacerbations, or fight infections
   c. Treat both chronic lung infections and acute pulmonary exacerbations
8. In people with CF, coughing is:
   a. Not a common problem
   b. How the disease is spread to others
   c. Hard on the lungs, and should be treated with cough medicines
   d. One way the body removes mucus from the lungs, and should be encouraged

Answers

To learn more, turn to the page number shown after the answer.

1. A, B, C
All of the statements about lung disease in CF are true. **CF affects the lungs. When breathing problems are first noticed is different for each person.** Page 41.

2. D
The function of mucus in the lower airways is to **trap inhaled particles and germs so they can be carried toward the throat and coughed up or swallowed.** The problem in CF is that the mucus is so thick and sticky that it cannot be easily carried to the throat, so the normal cleaning function of the mucus is impaired. Page 42.

3. B, C, D
It is true that **mucous plugs block the flow of air, if not removed can lead to lung infections and make it hard to remove particles and germs from the airways.** Page 42.

4. E
Any of the above (tiredness, weight loss and/or poor appetite, more coughing and/or wheezing, more mucus) may be symptoms of a pulmonary exacerbation. Page 44.

5. A, C
The purpose of airway clearance techniques (ACTs) in CF is to **clear the airways** and **loosen the mucus so it can be coughed out of the lungs.** ACTs help people with CF stay healthy. ACTs do not cure CF. Page 46-48.

6. A, B, C
For people with CF, it is true that **exercise helps to loosen mucus in the lungs, strengthen breathing muscles and may cause coughing and help clear the airways.** Exercise should NOT be avoided. Exercise is good for people with CF. Page 49.

7. C
In CF, antibiotics are used to **treat both chronic lung infections and acute pulmonary exacerbations.** Page 50.

8. D
In people with CF, coughing is **one way the body helps clear the lungs, and should be encouraged.** Page 46.

Notes and Questions

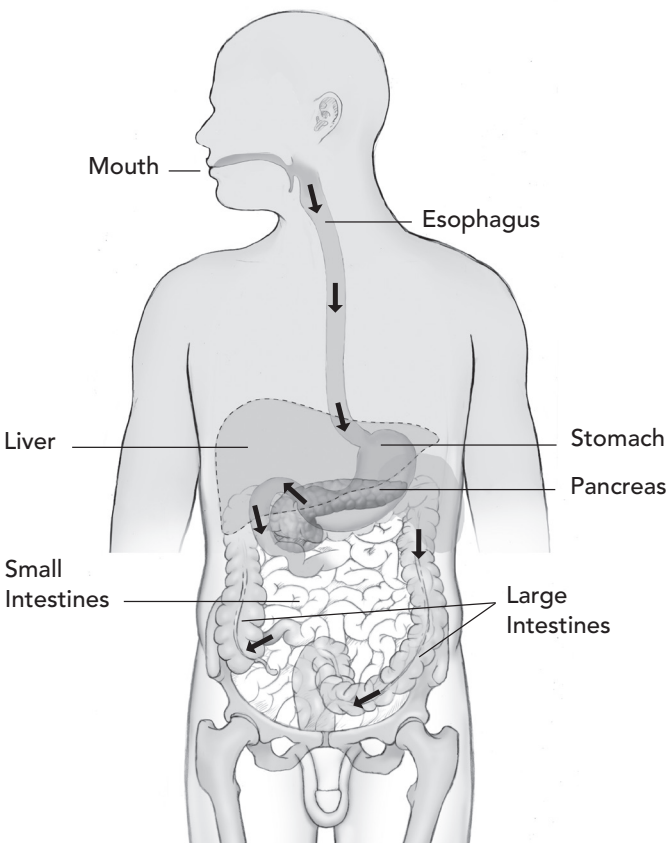
Chapter 5: Gastrointestinal System

Poor digestion is a common problem in cystic fibrosis (CF). The good news is that proper diet and enzyme supplements with meals and snacks can help solve this problem.

How the Gastrointestinal (GI) System Works

Digestion and food ***absorption*** are complex. When food is chewed and swallowed, it goes through the ***esophagus*** and into the stomach. Food in the stomach causes exocrine glands in the pancreas to make digestive enzymes. The pancreas is a gland in the body connected to the small intestine. Food goes from the stomach into the small intestine. The small intestine is where most food digestion and absorption occurs. Digestive enzymes from the pancreas enter the small intestine through a small duct, or passageway, called the ***pancreatic*** duct.

Other exocrine glands in the walls of the intestines make mucus to lubricate the digestive tract. This makes it easier for food and waste material to pass through the intestines.

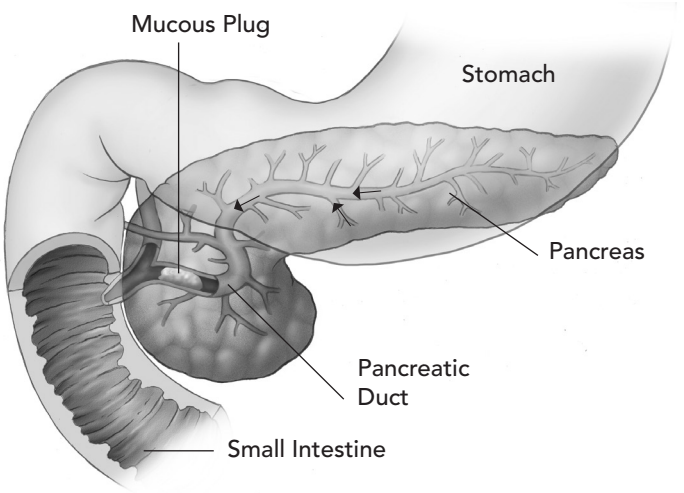


How the Gastrointestinal System Works

- *After food is swallowed, it goes down the esophagus and into the stomach. Food is churned in the stomach. Then, it moves into the small intestine.*
- *The pancreas makes enzymes to help digest fat and protein in foods. These enzymes enter the small intestine to digest food.*
- *What remains moves through the large intestine, where water is absorbed. Waste then moves out of the body.*

The Problem

For most people with CF, the exocrine glands in the pancreas make such thick secretions that the digestive enzymes are not able to get through the pancreatic ducts and do not reach the small intestine.



Digestive enzymes from the pancreas are blocked and do not make it into the small intestine.

With no enzymes to break down food, the food is poorly digested and absorbed. Much of the proteins, fats and carbohydrates in food are not absorbed for use in the body. This is called ***malabsorption***.

Malabsorption of proteins and fats can lead to poor growth and malnutrition. Proteins are needed for growth and body tissue repair or healing. Fats are calorie-rich food sources and give the energy needed for growth and development and to stay healthy. Fat also is needed for absorption of some ***vitamins***.

For some people with CF, the mucus that lubricates the intestines is so thick and sticky that it may block them. A blocked intestine needs special treatment.

Symptoms of Malabsorption

Symptoms of malabsorption include:

- Poor weight gain
- Frequent, large, greasy, foul-smelling stools
- Stomachache
- Excessive gas

Poor Weight Gain

Poor weight gain may occur even though the person with CF is hungry and eating a lot. It is caused by malabsorption of protein and fat. Lung disease may make weight gain even harder, because more calories are used for breathing and lung repair or healing.

Frequent, Large, Greasy, Foul-Smelling Stools

This is caused by the fat in the stools, because the fat is not absorbed for use by the body.

Stomachache or Abdominal Pain

People with malabsorption often complain of a stomachache, usually from poorly digested food in the intestine. This should

stop with treatment of malabsorption. At diagnosis, many parents of young children with CF report fussiness with feedings. This is likely because of discomfort from fat malabsorption. After a person with CF starts pancreatic enzyme replacements, this discomfort should go away. Other causes of stomachache are sore muscles from coughing or blocked intestines.

Excessive Gas

This also is caused by malabsorption.

People with CF who are not taking enzymes may have any or all of these symptoms. Once enzymes are started, the symptoms should improve. As children grow and gain weight, their dosage of enzymes will need to be increased. If children or adults who are taking enzymes develop symptoms of malabsorption, they may need an adjustment in the dosage of enzymes. Call your CF care provider to report symptoms of malabsorption. Remember not to change your dosage of enzymes without instruction from your CF care provider.

Treatment

For most people, the digestive, or ***gastrointestinal***, problems of CF are easy to treat. There are three common treatments:

- Diet and nutrition
- Pancreatic enzymes
- Vitamins

Diet and Nutrition

A good diet is important for normal growth and development. It helps the body fight infection. It helps lungs grow and heal. Good nutrition helps keep breathing muscles strong. Good nutrition has been shown to be closely related to good lung health in people with CF.

People with CF need the same good foods that everyone needs. In addition, they need more calories because of malabsorption. They also use more calories for breathing and to keep their lungs healthy.

People with CF may need up to **twice** the amount of calories per day compared with other people their age and weight.

For people with CF, fats are good! Fats are rich in calories and a good source of fuel for growth, development and energy. People with CF should eat a high-calorie, high-protein, high-fat, high-salt diet. Fats should not be restricted!

Most people with CF do well eating a healthy diet and taking pancreatic enzymes. If children are not gaining weight well, or adults are not maintaining a healthy weight, ask the CF doctor

or dietitian about ways to add calories. Nutritional supplements and ways to boost calories may help with weight gain. Besides high-calorie meals, snacks and nutritional supplements, some people with CF may need tube feedings to keep a healthy weight.

Talk to the CF doctor and dietitian about how a person with CF can use good nutrition to stay healthy.



Pancreatic enzymes come in capsules and are taken by mouth. They pass through the stomach and into the intestines, where they help to break down food so it can be absorbed by the body.

Pancreatic Enzyme Replacements or "Enzymes"

About 90 percent of people with CF need pancreatic enzymes to replace the natural enzymes that are not able to get through the blocked pancreatic duct. Enzyme replacements come in capsules and are taken by mouth. Inside each capsule are small beads that contain digestive enzymes. Each bead has a coating called an "enteric coating" that allows it to dissolve in the small intestine. The enzymes are then released in the small intestine to help digest food so the body can absorb it.

The amount (or dosage) of enzymes needed will vary with the person's age and weight, what he or she eats and how much CF has affected the pancreas. Enough enzymes should be taken to control symptoms of malabsorption. With enough enzymes, children should grow well, and adults should maintain a healthy weight.

Stomach acid may make pancreatic enzymes not work well. Sometimes acid blockers or antacids are prescribed to neutralize stomach acid.

Very high doses of enzymes can damage the large intestine. The CF doctor and dietitian will keep track of enzyme doses. If signs of malabsorption are present, call the CF care team. DO NOT give more enzymes on your own. Talk with the CF dietitian or doctor before giving more or less enzymes.

Pancreatic enzymes:

- Help digest fats, proteins and carbohydrates (the three parts of food that give us calories).
- Promote good nutrition and normal weight gain in children, and a healthy weight in adults.
- Help the body to absorb many essential nutrients.

How Enzymes Are Given

Enzymes should be taken at the beginning of each meal and snack. For infants and small children, the capsules may be opened. The beads in each capsule may be mixed in a small amount of soft acidic food, such as applesauce, and given from a spoon. For babies, it may help to give breast milk or formula after the beads are given, so that the beads are swallowed. Older children and adults can swallow the capsules.

More About Enzymes

If you notice that you are not getting the brand of enzymes your CF doctor ordered or that you usually take, contact your CF care center. They will help you to get the right enzymes.

- Enzymes should be taken at the beginning of each meal, snack or feeding.
- Take enzymes with all meals, snacks, milk, formula and nutritional supplements, including tube feedings that contain fat or protein.
- Do not skip taking enzymes. Keep some enzymes with you for meals or snacks away from home.
- Enzymes work for about an hour after taking them.
- Always take the right dosage, as prescribed by your CF care provider.
- Slightly higher dosages of enzymes may be needed with high-fat foods such as "fast foods," fried foods and pizza. Tell your CF dietitian or care provider if high-fat foods cause signs of malabsorption.
- Some foods and drinks do not require enzymes, because they contain only "simple carbohydrates" (or sugars) that digest easily. Examples of foods that do not require enzymes are fruits, juice, soft drinks, sports drinks, tea and coffee (without cream), hard candy (such as lollipops), fruit snacks, jelly beans, gum, popsicles or flavored ice.
- If the enzyme capsules are opened, do not crush or chew the beads. If beads are crushed or chewed, they will not work as well and could cause mouth sores.
- For infants and small children who need the capsules opened, mix the beads with a soft, acidic food, such as applesauce. Do not mix enzymes with milk-based foods such as yogurt or pudding because they will not work as well. Do not mix enzymes into foods ahead of time.
- Keep enzymes at room temperature (59° F to 86° F). Keep enzymes away from heat and cold. Do not store them in places that get hot or cold such as the top of a toaster oven, inside a hot car or in the refrigerator.
- Check the expiration date on each enzyme bottle to make sure they are "fresh."
- Ask the pharmacist if you can get the enzymes in the original, sealed, unopened bottle from the manufacturer.
- Always keep the enzyme bottle lid tightly sealed.



Most people with CF take enzymes before eating.

- Some babies, children, teens and even adults refuse to take their enzymes at times. Enzymes are essential in the care of CF. Talk with your CF care provider or dietitian if taking enzymes becomes a problem.

Vitamins

Because people with CF have trouble absorbing fats, certain vitamins are not absorbed from food. Vitamins A, D, E and K are "fat-soluble" vitamins. These vitamins need fat to be absorbed. Taking specially formulated water-soluble vitamins recommended by the CF care provider should help prevent vitamin deficiencies. These specially formulated vitamins should be taken once or twice daily, as prescribed by your CF care provider. The vitamins come in liquid, chewable tablet or capsule form. Chewable tablets should be chewed before they are swallowed.

Possible Gastrointestinal (GI) Complications

Meconium Ileus

For a few infants, the first symptom of CF is a blocked intestine at birth called meconium ileus. ***Meconium*** (***intestinal*** secretions) is normally passed by infants in their first 24 hours. About 1 baby in 5 with CF has meconium that is too thick. It blocks the intestines and has to be removed with enemas or surgery.

Distal Intestinal Obstructive Syndrome

Distal intestinal obstructive syndrome (DIOS) used to be called ***meconium ileus equivalent***. It is like meconium ileus, but occurs after infancy. It is caused by abnormal mucus in the intestine and poorly digested food. This results in stool that cannot move through the intestines normally. Partial or complete intestinal blockage may occur.

Causes of DIOS may include dehydration, not enough pancreatic enzymes and diet changes. The symptoms of DIOS are stomachache and fewer bowel movements than normal. If a person with CF has no bowel movements for 24 hours and has a stomachache, the CF care center should be called.

DIOS is often treated with:

- Good diet and enzyme replacement; sometimes the person will need to have nothing to eat for a period of time.
- More fluids.
- Special liquids to drink that move fluid into the intestine and stool — these liquids may be given through a nasogastric (NG) tube that is placed in the nose going into the stomach.
- Enemas.

DIOS must be treated promptly to prevent worse problems. It often can be treated with medicines, enemas and diet. Surgery is rarely needed to fix the obstruction.

Rectal Prolapse

Rectal prolapse is when the ***rectum*** sticks out through the anus. It is usually caused by hard-to-pass, abnormal stools, plus increased coughing and poor weight gain. It usually improves when food absorption and lung health improve. Surgery is rarely needed.

Other GI Problems

Cirrhosis of the Liver

The liver has ducts (or tubes) that drain ***bile*** secretions. These are called bile ducts. In CF, thick secretions may obstruct these ducts. This may cause ***cirrhosis*** of the liver. To watch for liver problems, CF doctors get a blood sample to check liver function yearly, and more often if needed. People with abnormal results on their liver function tests may be treated with medicines such as ursodeoxycholic acid (URSO). Serious liver problems are not common in CF. About 10 percent of people with CF have CF-related liver problems.

Cystic Fibrosis–Related Diabetes

Cystic fibrosis–related diabetes (CFRD) is a form of diabetes that can occur in people with CF. Diabetes is a problem in which a person's blood ***glucose*** (a type of sugar) level is too high. CFRD is not the same as diabetes in people without CF. The diagnosis and treatment are not exactly the same. If you or your child has been diagnosed with CFRD, you should be able to do all the things you want to do, and even eat all the foods you like. CFRD is common in people with CF especially as they get older. CFRD is found in about 35 percent of adults with CF ages 20 to 29 years and in about 43 percent of those ages 30 years and older.

Causes of CFRD

In people who do not have CF, the most common types of diabetes are type 1 and type 2 diabetes. Type 1 diabetes used to be called ***insulin***-dependent or juvenile-onset diabetes. It occurs most often in childhood. People with type 1 diabetes can't make any insulin, so they must take insulin to stay alive.

Type 2 diabetes used to be called non–insulin-dependent or adult-onset diabetes. It is caused by the lack of a normal response to insulin (sometimes called insulin-resistance) in addition to the body not making enough insulin. This type of diabetes occurs most often in people who are overweight.

People with type 2 diabetes do not always need insulin to manage their diabetes. Some take pills. Some use insulin. Some manage type 2 diabetes through diet, weight loss and exercise.

CFRD is different from type 1 or type 2 diabetes, but has some features of both. People with CFRD do not make enough insulin. This is probably because the thick mucus in the pancreas causes scarring. The scarring results in the pancreas not making enough insulin. Like people with type 2 diabetes, people with CFRD also have some insulin resistance. People with CFRD do not have diabetes because of being overweight. Therefore, unlike people with type 1 or 2 diabetes, people with CFRD should not treat their diabetes by limiting calories or losing weight.

Symptoms of CFRD

Having to urinate often (polyuria) and needing to drink often (polydipsia) are classic symptoms of diabetes. These symptoms are caused by high blood sugar levels. It is not easy to notice these symptoms in CF. People with CF often drink more (and then use the bathroom more). Other symptoms of CFRD include feeling very tired, losing or not gaining weight and unexplained decline in lung function. However, many people with CFRD do not have any symptoms at all.

CFRD Screening and Diagnosis

Because many people with CFRD do not know they have it until they are tested, your CF care provider will be testing you or your child for diabetes every year. Beginning at age 10 years, you or your child will have an ***oral glucose tolerance test (OGTT)***. An OGTT is done after a person has had nothing to eat or drink for 8 to 12 hours. Blood samples are taken before and up to 2 hours after drinking a set amount of glucose. The OGTT may be done sooner than 10 years of age if a person is having any symptoms of diabetes.

Treatment of CFRD

The treatment for CFRD is insulin. Insulin helps sugars and proteins to move from the blood into the body's cells. This is needed to provide energy and to build muscle. Insulin is given by injection. Some people with CFRD wish they could take a pill, like some people with type 2 diabetes do. Right now, only insulin is used to treat CFRD.

It is also important for people with CFRD to get enough calories. To ensure good health, it is important to maintain a healthy body weight. With type 1 or type 2 diabetes, people are often advised to eat a low-fat, low-salt and sometimes low-calorie diet. People with CF have different nutrition needs, though. When people have CFRD, they still need to eat their normal high-calorie,

high-protein, high-fat, high-salt diet to help get and maintain a healthy body weight. CFRD is not treated by limiting calories or losing weight.

Keeping blood glucose levels at a normal or near-normal level helps people with CFRD gain weight, feel better and have more energy. It also lowers the risk of problems caused by diabetes.

If you or your child is diagnosed with CFRD, your CF care provider will help you learn more about CFRD. He or she will also refer you to an ***endocrinologist*** who is a doctor with special training in the diagnosis and treatment of diabetes. Besides routine visits to the CF Foundation–accredited care center, people with CFRD should also be seen by a diabetes care team every 3 to 4 months. These visits are very important to help manage CFRD. You can also learn more about CFRD from the CF Foundation website at **www.cff.org**.

Gallstones

Gallstones occur in about 12 percent of people with CF. We do not know why. It may be related to how the body handles fats and bile acids. Gallstones do not often cause symptoms or need treatment. Rarely, surgery is needed to remove the ***gallbladder***.

Remember

Most of the gastrointestinal problems with CF are caused by blocked pancreatic ducts. The blockage keeps enzymes from reaching the small intestine. This causes digestive problems and malabsorption. But good, effective treatment is available. It involves:

- Pancreatic enzyme replacements — capsules/beads taken with food and drinks (such as milk or formula)
- Good nutrition — with foods high in calories, fats, proteins and salt
- Extra vitamins — specially formulated water-soluble vitamins
- Good fluid intake

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **The most common gastrointestinal problem in CF is:**
   a. Bile duct blockage that prevents bile from draining out of the liver
   b. Blockage in the bowel caused by thick secretions and abnormal stools
   c. Blockage in the pancreatic duct that prevents digestive enzymes from reaching the small intestine

2. **The blockage of enzymes in the pancreas can cause:**
   a. Ulcers in the bowel
   b. Poor digestion of protein and fat
   c. Excessive weight gain

3. **Symptoms of malabsorption in CF include:**
   a. Excessive gas
   b. Poor weight gain
   c. Stomachache
   d. Frequent, large, greasy stools
   e. All of the above symptoms
   f. None of the above symptoms

4. **These are all common treatments for malabsorption except:**
   a. Vitamins
   b A well-balanced diet
   c. A low-fat diet
   d. Pancreatic enzymes

5. **Which of the following statements is true about the use of pancreatic enzymes?**
   a. Enzymes should be taken with meals.
   b. Slightly higher doses of enzymes may be needed with high-fat foods.
   c. If the capsules are opened up, do not crush or chew the beads because they will not work as well.

Answers

To learn more, turn to the page number shown after the answer.

1. C
The most common gastrointestinal problem in CF is **blockage in the pancreatic duct that prevents digestive enzymes from reaching the small intestine.** Page 62.

2. B
The blockage of enzymes in the pancreas can cause **poor digestion of protein and fat**. This condition is called malabsorption. Page 62.

3. E
Symptoms of malabsorption in CF include **all of the above symptoms**. Excessive gas, poor weight gain, stomachache and frequent, large, greasy stools are all symptoms of malabsorption in CF. Page 62.

4. C
These are all common treatments of malabsorption except a **low-fat diet**. The poor digestion in many people with CF causes malabsorption of proteins, fats and carbohydrates. Pancreatic enzymes help digestion to occur more normally. A well-balanced diet high in calories, fats and proteins helps provide good nutrition for healthy growth and development. Pages 63-66.

5. A, B, C
All of these statements are true. Correct use of enzymes is essential to ensure that people with CF get all possible nutrients from the food they eat.
Pages 64-65.

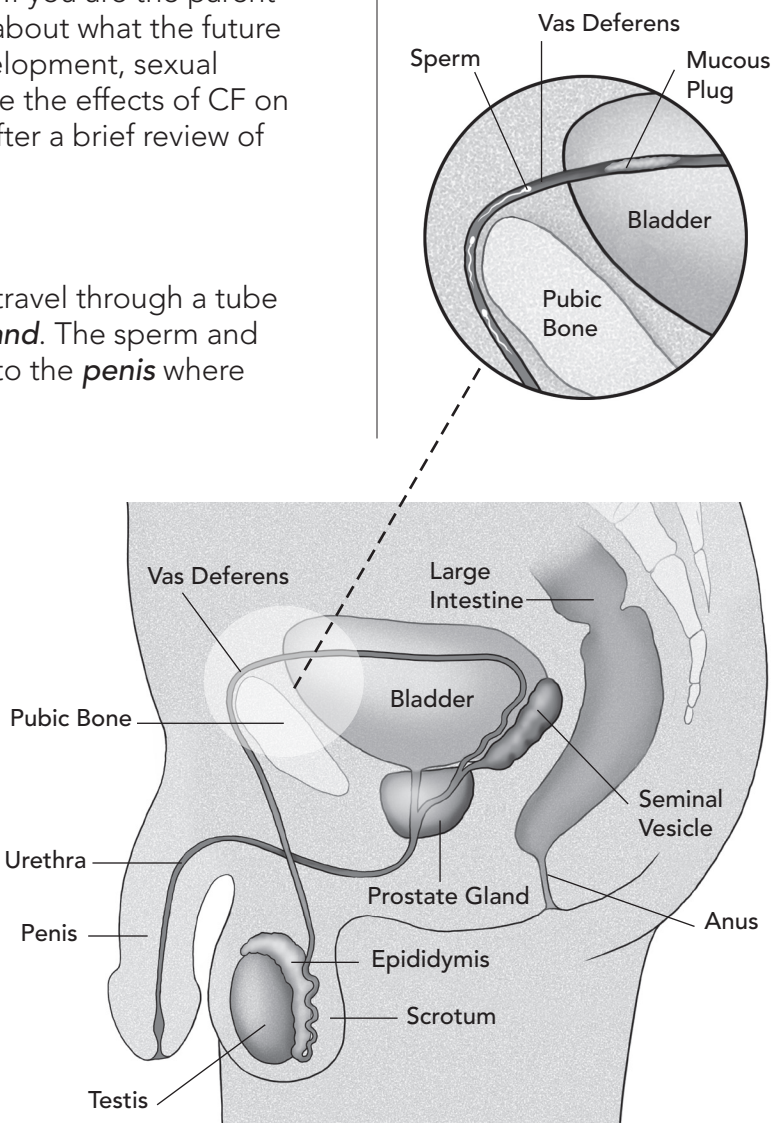
Notes and Questions

Reproductive System

Sexuality and reproduction are important concerns for teens and adults with and without cystic fibrosis (CF). If you are the parent of a young child, you may have questions about what the future holds for your child in terms of sexual development, sexual function and reproduction. We will describe the effects of CF on the reproductive system (the sex organs) after a brief review of normal function.

Male Reproductive System

Sperm are made in the ***testes***. The sperm travel through a tube called the ***vas deferens*** to the ***prostate gland***. The sperm and ***semen*** mix and travel through the urethra to the ***penis*** where they are ejaculated.



The Problem

In most males with CF, the vas deferens may not have developed normally or thick mucus may block it. Sperm cannot pass. Even though the testes make sperm and intercourse is normal, the semen ejaculated has no sperm so it cannot cause pregnancy. Men with CF are not ***sterile***, although most are ***infertile***. The sex hormones and glands are not affected by CF. Sexual development is normal. Sexual desire and performance are normal.

Absence of the vas deferens or blockage often occurs before birth. There is no way to fix this right now. Because of this, about 98 percent of males with CF are infertile. But men with CF should not assume they are infertile. In a few men with CF, the vas deferens is there and is not blocked. These men have some sperm in their semen and may be able to have children through intercourse.

A test, called a semen analysis (or sperm count), can be done when the male with CF matures. This test shows if sperm are in the semen.

Infertility Treatments for Men with CF

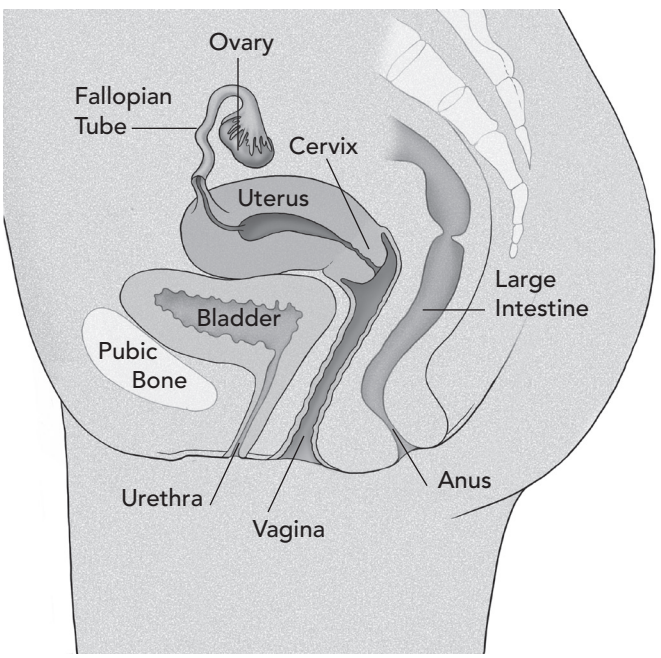
New infertility treatments mean that some men with CF can biologically father children. Sometimes, sperm can be taken from the testes, combined with an egg taken from the woman, and then the ***fertilized*** egg is put back into the woman. There are different procedures used to take sperm from males. These procedures are called:

- *Microepididymal sperm aspiration (MESA)*
- *Percutaneous epididymal sperm aspiration (PESA)*
- *Testicular sperm extraction (TESE)*

These procedures take sperm directly from the ***epididymis*** (fine tubules behind the testes). The procedure to collect the sperm involves an operation, usually done under local anesthesia.

After the sperm are collected, they can be used for in vitro fertilization, with or without ***intracytoplasmic sperm injection (ICSI)***. In vitro fertilization is a process where the egg cell is fertilized by the sperm outside the body. When ICSI is used, a single sperm is injected directly into the egg. The fertilized egg is then put into the woman's uterus.

Female Reproductive System



Eggs are made in the ***ovaries*** and go down the ***fallopian tubes*** to the ***uterus***, where the baby grows. Sperm, after being deposited in the ***vagina***, travel through the ***cervix*** and into the uterus to fertilize the egg. The vagina has exocrine glands that secrete mucus to lubricate the vagina and aid the passage of sperm. Women with CF may be less ***fertile*** than other women. However, conception is possible and many women with CF have had children.

The sex hormones and glands in women with CF are not affected. Sexual development is normal. Sexual desire and performance are normal.

The Problem

In women with CF, the vaginal and ***cervical*** mucus may be so thick and sticky that it makes it more difficult for the sperm to travel and reach the egg for ***fertilization*** to take place. Women with CF may ***ovulate*** (release eggs from the ovaries) less often and may have irregular periods, especially when having lung and/or nutrition problems.

Infertility Treatments for Women with CF

If a woman with CF makes the decision to have a child (see Making the Decision to Have a Child and Pregnancy, pages 76-77), she may be able to conceive without any infertility treatments. If infertility treatments are necessary, a fertility expert can perform a variety of tests to determine the exact causes of infertility. Some of the following treatments may be appropriate for a woman with CF who is trying to get pregnant.

Insemination

Insemination is a procedure when sperm are inserted into the cervix or directly into the uterus. Ovarian stimulation to produce more eggs may or may not be used with insemination.

In Vitro Fertilization

In vitro fertilization involves four steps:

1. Stimulation of the ovaries
2. Egg collection
3. Fertilization
4. ***Embryo*** transfer

Stimulation of the ovaries involves taking medication (hormones) to produce more eggs. Maturity of the eggs is determined by ultrasound and blood tests. Once the egg cells are mature, they are collected.

The mature eggs are collected under ultrasound guidance, with local anesthesia. The male partner is asked to produce a sperm sample on the same day. Fertilization takes place in the laboratory. The procedure is intended to produce a fertilized egg, called an embryo.

Embryos are usually transferred to the woman's uterus 48 hours after egg collection. Once the embryos are transferred to the uterus, medication is usually taken for 15 days to increase the chances of the embryo implanting in the uterus. A pregnancy test is then performed.

Safe Sex and Contraception

Most men with CF are infertile. Some women with CF are less likely to get pregnant. However, having CF is not birth control. All sexually active men and women with CF should practice safe sex. Protection from ***sexually transmitted diseases (STDs)*** is essential for everyone. Birth control should be used until a decision is made to have a child.

For people with CF, issues of ***contraception*** are similar for people who do not have CF, but there are some differences. Some forms of contraception are discussed in the next paragraphs. People with CF should talk to their CF care team and primary care physician or ***gynecologist*** about safe sex and which form of contraception is right for them.

The Condom

The condom provides contraception and protection from STDs.

Birth Control Pills

Birth control pills are often a good choice of contraception for women with CF. The pill does not provide any protection against STDs. Birth control pills may be less effective when certain antibiotics are taken. Also, women with liver disease or CFRD (diabetes) should talk to their gynecologist and CF doctor before starting birth control pills.

The Patch

The patch is a once-a-week birth control method that is as effective and works on the same principle as the pill. It does not give protection against STDs. When certain antibiotics are taken, the patch may be less effective. Some women like the patch better than the pill because it only has to be replaced once a week.

The IUD

An IUD is an intrauterine device made of plastic and/or copper that is inserted into the womb (uterus) by way of the vagina. There are different types of IUDs. Certain IUDs may be a good choice of contraception for a woman with CF. It does not give protection against STDs.

Contraceptive Injections

Contraceptive injections, such as Depo-Provera,™ are given by an injection every 3 months. It does not give protection against STDs. Contraception injections are NOT recommended for women with CF. People with CF are already susceptible to osteoporosis, a condition that results in bone loss. Using contraceptive injections further increases the risk of developing osteoporosis.

Making the Decision to Have a Child

For both men and women with CF, the question is not "Can I have child?" but "Is having a child right for me?" Like people who do not have CF, it is important for people with CF to make having a child a decision. Having a child is a life-changing,

long-term responsibility for all parents. People with CF who are making the decision about having a child should talk very openly to their CF care providers and obstetrician.

People with CF who are thinking about having a child should have genetic counseling. CF carrier screening should be offered to their partners.

Will a Parent with CF Have Children with CF?

Parents with CF will pass on one CF gene to all of their children. Because the parent with CF has two CF gene mutations, every sperm or egg will carry one CF gene mutation. So the genetic makeup of the partner will determine whether any children have CF.

If one parent has CF and the other does not have CF and is not a carrier of a CF gene mutation, then all children will be carriers. The children will not have CF.

If one parent has CF and the other is a CF carrier, there is a 50 percent chance that any child born will have CF. If the child does not have CF, the child will be a carrier of CF.

If both parents have CF, then any child they have together will have CF. Chapter 1 (pages 14-15) has more about how CF is inherited and the odds of having a child with CF.

Pregnancy

Women with CF should have good lung health and weight before getting pregnant. Having a healthy baby means making sure that the mother is healthy too. Many things affect how well a woman does with her pregnancy. CF affects pregnancy. Pregnancy affects CF.

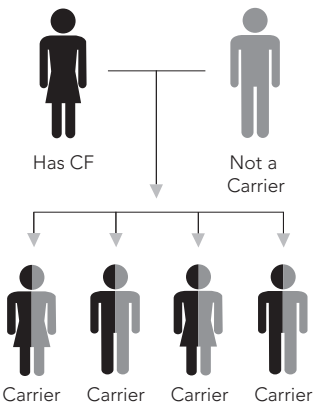
Women with CF can get pregnant. In the United States, hundreds of women with CF have had babies. How the mother and baby do during pregnancy depends primarily on the mother's health when she gets pregnant.

Any woman with CF who wants to get pregnant should know the possible risks. Also, the added demands of raising a child and how it affects her health should be considered. Because problems caused by CF vary, the decision is personal and should be discussed with her partner and a doctor who knows about CF and the woman's health.

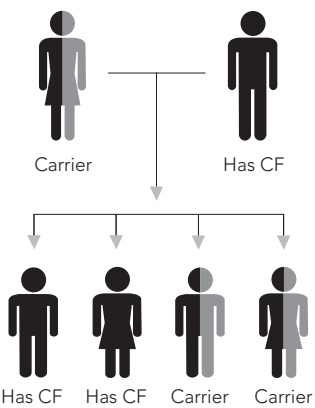
During pregnancy, women with CF should be very closely monitored by their CF care provider and high-risk obstetrician. They should be watched closely and treated early for pulmonary exacerbations. They should be monitored for diabetes. Careful attention should be paid to being well nourished and having good weight gain during pregnancy.

How a Person Gets CF

To have CF, you must get one copy of the CF gene from each parent. If one parent has CF, and the other parent is not a CF carrier all the children will be carriers.



When one parent has CF and one parent is a carrier, there is a 50 percent chance of having a child with CF.



Lung Function

The woman's lung health and lung function are the most important factors that affect her health and the baby's health. Women should talk to their CF care providers and obstetrician about how healthy their lungs are, and how a pregnancy is likely to affect them.

Nutritional Status

Women with CF need to be well nourished. The woman will need to be able to take in enough calories for herself and the baby to grow while pregnant.

Folic Acid

One of the most important things a woman can do to help prevent serious birth defects in her baby is to get enough folic acid every day, especially before conception and during early pregnancy.

After the Baby Is Born

After the baby is born, any parent with CF will need to continue to set aside time to take care of their own health. They will need to plan with their spouse and family, before the baby is born, so that they have help in place to take care of the baby and do their CF treatments.

Remember

For both men and women with CF, the sex hormones and glands are not affected. Sexual desire and performance are normal. Safe sex should always be practiced by everyone. Most males with CF are infertile. But men with CF should NOT assume they are infertile. Women with CF are sometimes less fertile than women without CF. Until making the decision to have a child, all sexually active men and women with CF should use contraception.

For more information, talk to your CF care provider. Your care center or the CF Foundation also has more resources.

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **Which of the following statements about CF and the male reproductive system is true?**
   a. Men with CF are usually infertile because of blockage or absence of sperm passageways, but men should NOT assume they are infertile.
   b. CF does affect the male reproductive system.
   c. Men with CF are not sterile.
   d. All of the above.

2. **Which of the following statements about CF and the female reproductive system is true?**
   a. Women with CF cannot become pregnant.
   b. CF does not affect the female reproductive system.
   c. Women with CF are usually sterile because of low hormone levels.
   d. Thick, sticky mucus makes it difficult for sperm to reach the egg.

3. **Which of the following statements is true about birth control pills?**
   a. Women with liver disease or CFRD should discuss the use of the pill carefully with their gynecologist and CF physician.
   b. They may cause problems for women with CF.
   c. Certain antibiotics may cause birth control pills to be less effective.
   d. Birth control pills are the only acceptable method of birth control for women with CF.

4. **Which of the following statements about women with CF are true?**
   a. They may be less fertile.
   b. They can become pregnant.
   c. They cannot have children safely.
   d. All children born to a woman with CF will carry at least one CF gene mutation.

Answers

To learn more, turn to the page number shown after the answer.

1. D

All of the above. All of the statements are true. **Men with CF are usually infertile because of blockage or absence of sperm passageways, but men should NOT assume they are infertile. CF does affect the male reproductive system. Men with CF are not sterile.** Page 73.

2. D

It is true that thick, **sticky mucus makes it more difficult for sperm to reach the egg.** Page 74.

3. A, C

The following statements are true: **Women with liver disease or CFRD should discuss the use of the pill carefully with their gynecologist and CF doctor and certain antibiotics may cause birth control pills to be less effective.** A gynecologist who knows CF should be consulted before use of birth control pills. An alternative form of birth control should be used when the woman is on certain antibiotics. Page 76.

4. A, B, D

It is true that women with CF **may be less fertile, they can become pregnant and that all children born to a woman with CF will carry at least one CF gene mutation.** Women with CF can have a safe pregnancy and healthy children. How the mother with CF and baby do during pregnancy depends on the mother's health when she gets pregnant. A woman with CF should talk with her CF care team and partner before becoming pregnant. Pages 74-78.

Notes and Questions

PART III

Living with Cystic Fibrosis

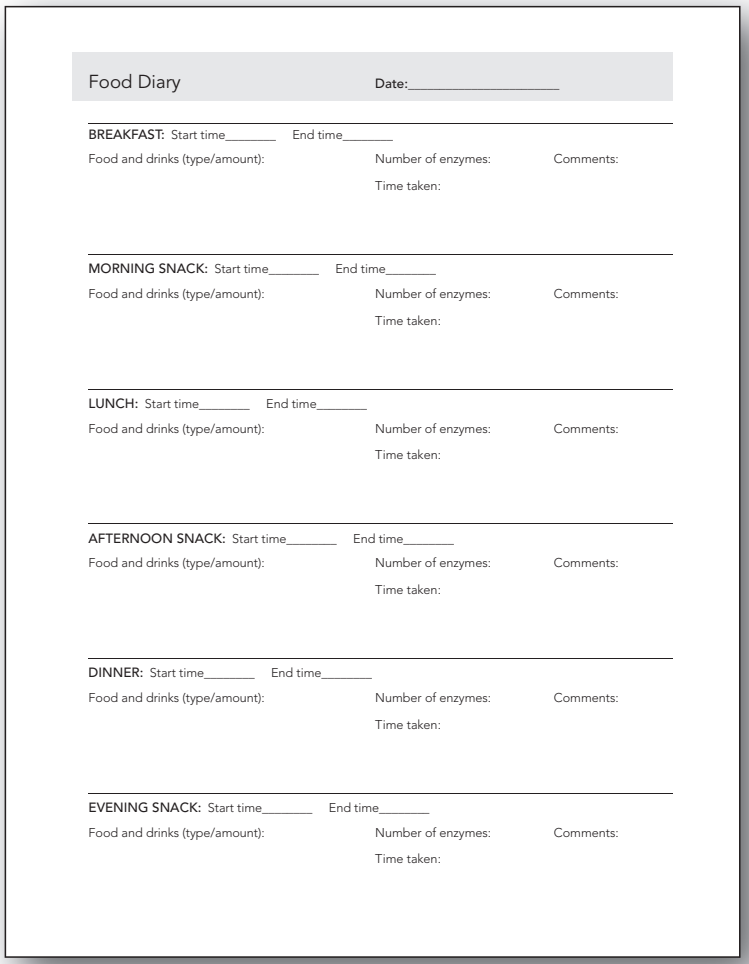


Overview: Living with CF

Life with cystic fibrosis (CF) may seem filled with a lot of schedules, treatments and diet rules. People with CF often face four main issues:

1. Staying fit and healthy
2. Sticking to daily treatments
3. Managing respiratory problems
4. Managing nutrition and digestive problems

Parents manage these issues for small children. They also teach their child to manage his or her own CF care as soon as the child is able. Start promoting independence early, so that by the time children are ages 12 to 14 years they do most of their own care. Teens should spend the first part of their clinic visits talking with the CF care team by themselves. The parent can then join them toward the end of the visit.



Food Diary Date:_____

BREAKFAST: Start time_____ End time_____
Food and drinks (type/amount): Number of enzymes: Comments:
Time taken:

MORNING SNACK: Start time_____ End time_____
Food and drinks (type/amount): Number of enzymes: Comments:
Time taken:

LUNCH: Start time_____ End time_____
Food and drinks (type/amount): Number of enzymes: Comments:
Time taken:

AFTERNOON SNACK: Start time_____ End time_____
Food and drinks (type/amount): Number of enzymes: Comments:
Time taken:

DINNER: Start time_____ End time_____
Food and drinks (type/amount): Number of enzymes: Comments:
Time taken:

EVENING SNACK: Start time_____ End time_____
Food and drinks (type/amount): Number of enzymes: Comments:
Time taken:

Some people find it helpful to make a daily calendar of CF care. This will help them keep track of what needs to be done and when. It may also be helpful to keep a diary of daily fluid and food intake, medicines, enzymes and bowel habits. Letting the CF doctor see it at the next checkup can help show, for example, if the enzyme dose is correct. Some people keep computer records of their medicines. This helps them keep track of prescription refills. Before going to the clinic, they can print updated lists to show the care team.

See Appendix D, page 195, for examples of daily record forms that can be used to keep a food diary, a treatment diary and a symptom diary. Your CF care center staff can also offer suggestions for sources for forms and programs to help you keep such records.

Notes and Questions

Chapter 8: Staying Healthy: What You Can Do

We have described cystic fibrosis (CF), how CF affects the body and treatments for CF. With routine therapies and regular visits to a Cystic Fibrosis Foundation–accredited care center, most people with CF can lead active lives. This chapter describes other things to do to stay healthy.

Knowing Yourself

Staying healthy begins with knowing yourself, your body and how you feel. Of course, for infants and children, parents need to observe their behavior and health symptoms.

Everyone, with or without CF, needs to "listen to their body." For example, our bodies tell us when we're hungry and need to eat, and when we're tired and need to rest. Listening to our bodies helps us know how to take care of ourselves. People with CF also need to know how CF affects their body, which will help them understand what their body is telling them.



Exercise strengthens muscles and increases appetite. Like everyone else, people with CF should exercise regularly.

For example, some people with CF have a slight cough that they may notice only in the morning when they wake up. For those people, when they notice a cough off and on during the day, they know that their body is telling them that something is going on — that their CF may be worsening a little. This is the time to call their CF care provider. It might be time to go in for a CF clinic visit, get some oral antibiotics and increase airway clearance from 2 times per day to 3 times per day for a week or two.

Another example of "knowing your CF" is for people with CF to know how energetic they feel when they are feeling their best. Sometimes people with CF just don't feel as energetic as they normally do. Sometimes having less of an appetite goes along with having less energy. These may be signs of an early lung infection or pulmonary exacerbation (even without having a cough!). This would be the time to call or see the CF care team.

Noticing changes in symptoms early and getting treatment early can help keep you or your child from having worse problems that are harder to treat.

Knowing how you or your child feels when feeling really well is also called knowing your or your child's "best baseline." Knowing their best baseline helps people with CF recognize when they are not feeling their best — that's the time to call or visit the CF care center.

When a person is feeling his or her best and healthy, he or she is energetic, active and interested in interacting with the world around them. All people should be able to have interests and hobbies and goals and dreams for what they want to do with their lives.

Based on your or your child's best baseline and goals, you and the rest of your CF care team will make a plan for you or your child to stay healthy, which we call the CF treatment plan. Treatment plans typically include a high-calorie, high-fat diet; airway clearance therapies to loosen the clogged mucus from the airways; aerosolized medications for the lungs; enzymes and vitamins for nutrition; and a plan to be active and exercise.

Sticking to Your Treatment Plan

Sticking to your treatment plan can be thought of in two ways:

1. Sticking to your or your child's plan of doing the prescribed treatments every day.
2. Sticking to a plan of making visits to your CF care center to see your team.

Sticking to the daily treatment plan helps to keep a person with CF active and healthy. Even though the person with CF may not actually "feel" it, these daily treatments are keeping his or her lungs healthy. It's important to find a way to make daily CF care a part of your routine daily activities.

Making regular visits to your CF care center is a very important part of the CF treatment plan. People with CF need to visit their CF care center at least four times per year for regular visits and all of the recommended tests. Some people have treatment plans that include regular visits more often than every 3 months. More visits are necessary when a person with CF is not feeling his or her best.

Why are CF care center visits important, even when a person with CF is feeling "okay"? Your CF care provider and team need to see people with CF regularly to see how they are doing and monitor their health on a regular basis, just as people that do not have CF need to see their ***primary care provider (PCP)*** on a regular basis.

For people who do not have CF, the PCP may find that they have blood pressure that is higher than normal or, on a routine blood test, a cholesterol level that is too high. High blood pressure and cholesterol level can be treated early and may prevent a stroke or heart attack! Yet when your blood pressure or cholesterol level start going up, you can't "feel" that! But your PCP can determine that there is a problem.

For people with CF, regular visits to the CF center include tests such as pulmonary function tests (PFTs), sputum or throat cultures and blood tests. People with CF cannot feel small decreases in lung function or a germ growing in the lungs. They only feel symptoms when these issues get worse and start causing bigger problems.

By the time a person with CF notices symptoms, he or she may have big decreases in lung function, more symptoms and even some permanent lung damage. Your CF care provider can detect these things early with tests and can treat these problems early. For example, early treatments of a first-time growth of *Pseudomonas* may be able to get rid of the germs from your lungs. If the *Pseudomonas* is not caught and treated early enough, a person with CF might have to deal with *Pseudomonas* for the rest of his or her life.

CF clinic visits also give you the opportunity to talk to your CF team about how you or your child is doing. Ongoing, open and honest communication with your CF care team will make it possible to continue adjusting the treatment plan to be the best for you or your child.

Daily care for CF takes a lot of time and commitment. We live in a busy world, where none of us seems to have enough time. Family and close friends can be a great support to you. And remember, you are part of a team, and team members help each other. Talk to your CF team members! Your CF team social worker and psychologist can help you, for example, with time management and coping skills. Sometimes it takes finding ways to stick to your treatment plan. Sometimes it takes adjusting your treatment plan. Your team is experienced in helping people with CF and their families with these issues, so don't hesitate to ask them for advice.

CF treatment plans change over time. Talk to your CF care team about how you or your child feels, and how you feel about the treatment plan. Sticking to your CF treatment plan may seem difficult at times. But when you take the time to eat well, exercise and be active; do your daily CF treatments and medications; and visit your CF care center regularly, you will find that you have more energy to do the things you want to do.

Exercising and Staying Active

Exercise and staying active should be a part of everyone's daily routine. People of all ages with CF, including those with lung disease, can and should exercise. People who have CF can benefit in many important ways from being active and exercising. Remember that exercise does not have to be something formal or done in a gym. It can be playing soccer with friends in the park.

There are so many benefits of exercise and staying active! Some are listed here:



People with CF should be physically active. Most people with CF have no limitations when it comes to exercising. In fact, it has many benefits for those with CF.

- Exercise helps keep lungs healthy.
- Exercise loosens mucus in the lungs so the mucus can be coughed up more easily.
- Exercise makes breathing deeper, and gets air around mucus in the airways.
- Exercise (such as lifting weights and doing push-ups) helps build stronger muscles.
- Stretching exercises can improve flexibility and posture. Good posture can make it easier for a person with CF to take deep breaths.
- Weight-bearing exercise (such as walking, jumping rope, playing basketball and running) helps strengthen bones and can prevent osteoporosis.
- Exercise can help improve a person's energy level.
- Exercise increases appetite. A person with a good appetite eats more food, is more likely to have better nutrition and is more likely to have more energy.
- Better fitness leads to longer and healthier lives for people with CF and without CF.
- Regular exercise promotes a feeling of well-being, decreases stress and improves mood.

People with CF should talk to their CF care provider or physical therapist about an exercise and activity program that is right for them. Remember that people with CF need extra fluids and salt with increased sweating or in hot weather.

Good Nutrition

Good nutrition is very important for people with CF of all ages. There is a connection between good lung health and good nutrition. A balanced, high-calorie diet with fat and protein gives the body what it needs to grow normally and live well. Remember: people with CF need extra calories to meet the needs of their bodies.

Good nutrition

- Develops strong lungs.
- Keeps lungs as healthy as possible.
- Builds and keeps a strong immune system to fight infections.

Getting Children to Eat

Getting children to eat can often be a challenge. For parents of a child with CF, this can be a real source of concern. Here are some things that parents can do to help children eat well to stay healthy.

Plan Ahead

- Have three meals and three scheduled snacks every day, at about the same time every day.
- Before starting a meal or snack, do your best to have all the food ready.
- Reduce distractions (no TV, toys, games or books at the table).

Make Mealtimes Structured

- Let your child know that it's time to eat. (Say, "It's breakfast time," rather than asking,"Would you like some breakfast now?")
- Do not coax and beg your children to eat or punish them for not eating.
- Sit at the table with your children. Relax and enjoy being together.
- Praise your children when they are eating well.
- Limit the length of meals to about 20 minutes.
- Reward your child for good mealtime behavior with a nonfood treat.

Give Choices

- Do not try to "trick" your child by hiding new or disliked foods in food they like.
- Let young children choose whether to open their enzyme capsules or have you do it for them.
- Allow your child to choose between two high-energy/high-calorie additives to foods (for example, extra cheese or extra butter or both).
- Even if your child chooses not to eat a new food, keep offering it. It often takes 10 to 15 "exposures" to new foods before children are willing to try them.

Dealing With Emotions and Attitudes

- Try not to worry if a meal ends and your child has not eaten much. He or she will eventually get hungry.
- Have a relaxed discussion with your child about the importance of healthy eating, at a time other than meals or in CF clinic.
- Give your child an opportunity to talk about his or her feelings about having to eat a lot and about having CF.
- Talk frequently with the dietitian, nurse, social worker and/or psychologist in your center for support and suggestions.
- Ask the dietitian, nurse, social worker and/or psychologist to connect you with another parent of a child with CF who has found things that helped improve his or her child's eating.
- For more information about getting children to eat, you may want to read *Nutrition: How to Encourage Healthy Eating*, which is available from your CF care center or the CF Foundation's website at **www.cff.org**.

Adolescence, Puberty and Nutrition

Adolescence, or the teenage years, is the time of transition from being a child to becoming an adult. Puberty, the period of time when a person's body changes and becomes sexually mature, takes place during adolescence. There is no other time in life, except during infancy, when there is such intense growth and development. Puberty is a process that takes 3 to 5 years.



At this time, for a young person with CF, getting enough calories is very important to support all of the physical growth and changes he or she is going through. Good nutrition and enough body fat will help an adolescent with CF start puberty and grow like their peers without CF.

Adults

For adults with CF, your busy work or school schedules can get in the way of preparing high-calorie meals and snacks. You can ask your CF dietitian about how you can make some quick-to-grab snacks and plan for meals. For helpful suggestions, visit the "Staying Healthy" section of the CF Foundation's website at **www.cff.org**.

Cystic Fibrosis–Related Diabetes (CFRD)

When people have cystic fibrosis-related diabetes (CFRD), it is important to keep it under control. Keeping the glucose at a normal or near-normal level and getting enough calories helps people with CFRD:

- Gain weight (for children) and maintain a healthy weight (for adults).
- Feel better and have more energy.
- Have better lung function and fewer pulmonary exacerbations.
- Lower their risk of long-term problems that diabetes can cause (including blindness and kidney disease).

Bone Health

As people get older, bones get weaker and can break more easily. Two common bone diseases are osteoporosis and *osteopenia*.



Minerals such as calcium, phosphorus, magnesium and fluoride build bones. With osteopenia, bones do not have enough of these minerals. This is called low mineral density, or low bone density. Normal bones look like a "honeycomb" and have holes in them. With osteoporosis, because of low bone density, the holes get too big and make the bones weak.

People who do not have CF are at risk for osteoporosis and osteopenia, but this is usually a problem that begins when people are in their 50s or older. People with CF are at risk for osteoporosis and osteopenia at a much younger age, even in their teens.

Things that help keep bones healthy include:

- Good nutrition: getting enough calories and nutrients, including calcium and vitamin D
- Taking enzymes and fat-soluble vitamins
- Weight-bearing and resistance exercise such as walking, jogging or weight lifting
- Getting regular CF care and getting tests to check nutrition (including vitamin levels) and special bone X-rays as your CF care provider and dietitian recommend
- Avoiding things that lower bone density, including alcohol, caffeine, carbonated drinks and smoking

For more information on bone health, you may want to read "*Nutrition: Bone Health and Cystic Fibrosis*," available from your CF care center or the CF Foundation at **www.cff.org**.

Ways to Increase Calories

People with CF need more calories than people without CF. This is true for children who are growing, and for adults who are trying to maintain a healthy weight while managing busy lives. Sometimes people with CF may need up to twice the calories that a person without CF their same age, sex, height and weight would need. Taking in this many calories can be hard to do. Even when people with CF eat often and well, they still may not be getting enough calories.

Some ways to add calories to the diet are:

- Drinking milkshakes and smoothies or nutrition drinks.
- Trying some fast and easy snacks and meals such as frozen breakfasts, sandwiches or frozen meals to microwave, cheese and cracker snacks, granola or protein bars, pudding snacks, nuts or sunflower seeds.

Talk to your CF dietitian for more ideas based on your lifestyle and what you like to eat!

Avoid Smoking and Secondhand Smoke

Breathing tobacco smoke is harmful to the lungs of all people — especially for those with CF. This includes secondhand smoke. Even the particles left behind on a smoker's clothes, skin, hair and breath can irritate the airways of people with CF.

Caregivers

Parents, relatives and caregivers of children with CF should not smoke. If parents smoke, their children are more likely to smoke. One of the best things you can do to help yourself or your child stay healthy is to have a smoke-free environment. If you smoke, there is help for quitting. Call the American Lung Association at **(800) LUNG-USA** or go to **www.lungusa.org**.

While you are working to quit smoking, here are some simple things you can do to help keep your child healthy:

- Smoke outside away from your child.
- Do not smoke in your home or car.
- Do not let others smoke around your child or in your home or car.

Secondhand Smoke and Lung Irritants

Secondhand smoke in enclosed areas, such as cars, homes, apartments and restaurants remains in enclosed areas for hours, days or longer. If a relative or friend who smokes in his or her home invites you over with the promise that they won't smoke while you are there, the secondhand smoke is still present in the air that you breathe.

People with CF also should not work in an environment where they are exposed to secondhand smoke, such as bars in cities or states that do not have laws against smoking in public places. They should also not work in places that expose them to other lung irritants, such as dust, paint fumes, volatile chemicals and vehicle exhaust.

Smoking by People with CF

For people with CF, smoking is disastrous. Quite simply, the effects of smoking damage your lungs' ability to breathe. For young people, smoking harms the lungs' ability to grow and reach their full potential. Exposure to tobacco smoke also makes people more likely to get respiratory tract illnesses, and for those illnesses to be more severe.

Tobacco smoke — even secondhand — irritates the lungs and sinuses and causes inflammation and swelling, which damages the tissue. People with CF already have a hard time eating enough and gaining weight. Exposure to tobacco smoke can decrease the sense of smell and taste, thus decreasing their appetites. Being a smoker may even disqualify a person from being eligible for a lung transplant.

Tobacco smoke also shuts down the cilia, the tiny hairlike cells on the lining of the airways that help remove mucus, dirt and germs from the lungs and sinuses. Compared with normal lungs, lungs with CF have mucus that is thicker, and the cilia have less fluid to help them move. When the cilia are weakened by smoke, they are even less able to keep the lungs clean.

When you have CF, you must constantly work to keep your lungs in good shape so you can live a long, healthy life. To put it bluntly, smoking and even exposure to other people's smoke will damage your lungs and make you sicker, and, ultimately, shorten your life.

Vaccinations and Avoiding Germs

It is always better to prevent a health problem than to try to fix one after it has started. For instance, vaccinations (immunizations) are vital for prevention. Everyone should get the recommended vaccinations, and a yearly flu shot. Avoiding germs as much as possible is always best. Washing hands or hand hygiene should be done routinely. Read more about avoiding germs in Chapter 9, *Staying Healthy: Avoiding Germs*.

Remember

There is a lot that you can do to help keep yourself or your child with CF healthy and prevent difficult problems from occurring. Know your or your child's usual health status so that when something changes, you will be ready to tackle it early, before it develops into something worse.

Sticking with the daily CF treatments may seem tiresome at times, but it is extremely important in order to keep a person with CF as healthy as possible. Good nutrition, including a high-calorie diet, is vital for a person with CF. Staying away from tobacco smoke, including secondhand smoke, will prevent lung damage. Although no one can avoid exposure to all germs, doing good hand hygiene and getting the recommended vaccinations will help a great deal. Your CF care team can help you find ways to fit all of this into your life.

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **Which of the following statements about tobacco smoke are true?**
   a. Exposure to tobacco smoke makes people more likely to have respiratory tract illnesses.
   b. If parents smoke, their children are more likely to smoke.
   c. Secondhand smoke can linger in an enclosed area for hours, days or longer.
   d. Being a smoker may disqualify a person from being eligible for a lung transplant.
   e. All of the above

2. **Which of the following are ways to get some exercise?**
   a. Playing soccer with friends on the weekend
   b. Being on the high school football team and participating in all practices and games
   c. Working out at the gym 3 days a week during lunch time
   d. Going to a family picnic and running around and playing games
   e. All of the above

Answers

To learn more, turn to the page number shown after the answer.

1. E

All of the above are true. Exposure to tobacco smoke makes people more likely to have respiratory tract illnesses. If parents smoke, their child is more likely to copy them and smoke. Secondhand smoke can remain in an enclosed area for hours, days or longer. Being a smoker may disqualify a person from being eligible for a lung transplant. Pages 94-95.

2. E

All of the above. There are many ways to get exercise. Exercise can be a formal schedule of walking, running or working out; participation in sports; and informal activities. Page 90.

Notes and Questions

Chapter 9: Staying Healthy: Avoiding Germs

We live in a germ-filled world. People with cystic fibrosis (CF) get more lung infections, so some things should be done to cut their risk. For the person with CF and his or her family, this means a balance between ignoring germs and worrying about them. It is best to help the body fight germs and avoid germs when possible.

Germs and CF

Germs are tiny organisms that can cause infection. Germs are all around us. For people with CF, some germs can cause serious lung problems.

There are three major groups of germs:

1. Bacteria
2. ***Viruses***
3. Fungi and molds

Many CF germs are spread by contact (or touch) and ***droplets*** from coughing, sneezing and nose blowing.

Some germs, such as the viruses that cause colds, the flu and ***respiratory syncytial virus (RSV)*** affect people with and without CF. But when people with CF get a respiratory virus, they may get sicker because of their lung disease. Other germs — such as the bacteria *Pseudomonas aeruginosa (Pseudomonas)* and ***Burkholderia cepacia complex*** *(B. cepacia)* — cause more harm in people with CF than in people who do not have CF.



To prevent the spread of germs, use a tissue when coughing and clean your hands afterward.

Remember that the gene mutation that causes CF leads to a problem in how salt moves in and out of the cells in the lungs. This causes thick, sticky mucus. Germs stick, stay and grow well in this mucus in the lungs. The airways swell because of inflammation and make more mucus. More germs grow, and the cycle continues.

How Do Germs Spread?

Germs, such as bacteria and viruses that cause lung infection, can spread between people in many ways. These are known as "routes of transmission."

The three main ways for germs to spread are:

1. By contact
2. In a droplet
3. Through tiny remains of droplets floating in air (airborne)

The most common way germs spread is by contact. This is also called "***direct contact transmission***" or "***indirect contact transmission.***" Viruses that cause common colds, RSV and CF-specific germs such as *Pseudomonas* and *B. cepacia* are spread this way.

Direct contact is when germs spread by bodies touching, such as through shaking hands, hugging or kissing.

Indirect contact involves touching something with germs on it, such as touching a doorknob or sharing a cup. Germs spread to you when you touch something with germs on it and then touch your eyes, nose or mouth.

When a person talks, sings, coughs, sneezes or laughs, droplets are made. These tiny drops of liquid may have germs inside. The droplets with germs can land in the eyes, noses or mouths of others. This is how germs can be spread by droplet transmission. These drops can travel 6 feet and more through the air before they fall to the ground. The flu (influenza) and whooping cough (pertussis) are spread this way.

Some germs travel through the air on specks of dust or particles made when a person talks, sings, sneezes, coughs or laughs. These germs can float in the air for a long time. They can be carried a long way by air currents. Illness occurs when people breathe in the germs floating in air. Tuberculosis (TB), measles and chickenpox are some of the germs spread by airborne transmission.

Routine Scheduled Vaccinations for Children:

- Diphtheria
- Haemophilus B (Hib)
- Hepatitis A
- Hepatitis B
- Human papillomavirus (HPV)
- Measles
- Meningococcal
- Mumps
- Pertussis
- Pneumococcal
- Polio
- Rotavirus
- Rubella
- Tetanus
- Varicella (chickenpox)

Vaccinations

One of the best ways to help the body fight germs is for people to get their vaccinations. Vaccinations are also known as immunizations, or "shots." There are 2 different groups of vaccinations: routine scheduled vaccinations, and seasonal or yearly/annual vaccinations.

Routine Scheduled Vaccinations

EVERYONE, children and adults, should get the vaccinations or shots recommended by the Centers for Disease Control and Prevention (CDC). These vaccines help prevent illness. Getting vaccinated, or immunized, is easy, low cost and saves lives!

All people with CF should get all their routine immunizations. Household members, close relatives and people in close contact with people with CF should also be immunized.

Children should get their first immunizations before they are 2 months old. Older children and adults need to get routine scheduled vaccinations throughout their lives. People should get their vaccinations from their primary care providers. They may also be able to give you information about special programs that provide free immunizations.

If a person is behind on routine immunizations, he or she should check with his or her primary care provider to schedule the vaccinations needed to get caught up.

Seasonal Vaccinations

Influenza (Flu)

Influenza (the flu) is an illness caused by a virus that can make people very sick. Every year in the United States, flu epidemics occur during the winter months. Influenza is very ***contagious***. It can spread from person to person in droplets created by coughing and sneezing. It can also be spread when people cough or sneeze onto their hands and then touch things.

Anyone can get the flu, including people with CF. However, when people with CF get the flu, they can get much sicker than people who do not have CF. In people with CF, the flu can lead to a lung infection.

Flu outbreaks in the United States usually occur between December and March. Flu shots are often available as early as August. People with CF should get the flu shot as soon as it is available.

People with CF who are 6 months and older, and who are not allergic to eggs, should get a flu shot every year in the fall. Household members, relatives, day care and health care staff and close friends of those with CF also should get flu vaccines to prevent the flu. The flu vaccine comes as a shot or nasal spray. People with CF should NOT get the nasal spray vaccine.

The flu vaccine does not give you the flu. Viruses in the flu shot are killed (inactivated). They are weakened (attenuated) in the nose spray. The risk of the flu vaccine causing serious harm is extremely small. However, like any vaccine, mild side effects are possible. Side effects may be a low-grade fever or some mild aches that may begin shortly after the vaccination and usually last 1 or 2 days. The shot may cause soreness, redness or swelling where it was given.

People who are allergic to eggs should talk to their primary care provider or CF care provider about other ways to prevent the flu.

Routine Scheduled Vaccinations for Adults:

- Diphtheria
- Hepatitis A
- Hepatitis B
- Human papillomavirus (HPV)
- Measles
- Meningococcal
- Mumps
- Pertussis
- Pneumococcal
- Rubella
- Tetanus
- Varicella (chickenpox)
- Zoster

Symptoms of the flu include:

- Body aches and headache
- Fatigue
- Fever and chills
- Increased cough
- Sore throat

If you or your child gets the flu, be sure to call your CF care center, even if you did get the flu vaccine. If you or your child did NOT get a flu vaccine and are exposed to someone with the flu, call your CF care center. Your CF care provider may recommend starting a medicine that may help you or your child have milder symptoms and a shorter illness if you do get the flu.

Pandemic Viruses

Every year some people get the flu in the fall and winter. That is why it is called "seasonal" flu. However, sometimes new influenza viruses appear. These situations are called pandemic, or worldwide infections. During pandemics of new flu viruses, people can get the flu at any time during the year.

In 2009, for example, there was a pandemic flu caused by the H1N1 flu virus. It was also called "swine flu." The virus was first detected in the United States in April 2009, and it was not limited to a specific season. Now, when you get your flu shot, you are also protected from the H1N1 flu virus.

Pandemic flu occurs from time to time. During these times, be sure to find out if you should receive another flu vaccine.

Respiratory Syncytial Virus (RSV)

Respiratory Syncytial Virus (RSV) is another germ that anyone can get. RSV can make babies, young children and the elderly very sick. There is a special shot to decrease how severe RSV is in young children. The child must receive the shot every month during RSV season, which is the winter months. The CF Foundation recommends that the RSV shot is considered for children with CF younger than 2 years old. Good hand cleaning reduces the risk of getting RSV or other germs for people of all ages.

Getting immunized is a very important way of avoiding germs. Everyone should get all their routine vaccinations and yearly flu vaccines and maybe vaccines for other viruses. Everyone should make sure to get all vaccinations recommended by the CDC at the proper time, and to keep records of all immunizations. For more and current recommended vaccinations, refer to the CDC. Call toll free **1-800-CDC-INFO** or **1-800-232-4636**, or visit **www.cdc.gov/vaccines**.

Handwashing (Hand Hygiene)



You can reduce the risk of germs by cleaning your hands regularly.

One of the best ways to keep from catching or spreading germs is handwashing, or ***hand hygiene***. Clean your hands often. (See "When to Clean Your Hands" in the next section.) Clean your hands after touching things that have contacted ***mucous membranes***, sputum or phlegm.

There are two ways to clean your hands. The first is to clean your hands with soap and water, the second is to clean your hands with alcohol-based hand gel (see "Cleaning with Soap and Water" and "Cleaning with Hand Gel.")

If you see dirt on your hands, wash with soap and water. If you do not see dirt, you can wash with soap and water, or use alcohol-based hand gel. Hand gel or sanitizer should contain at least 60 percent alcohol to clean hands. Some people carry hand gel with them to use before eating and after nose blowing, coughing, sneezing or shaking hands.

People with CF, parents and siblings can reduce some of the risks of illness by regular handwashing or hand hygiene. Good handwashing or hand hygiene should be practiced everywhere — at home, school and work, during play, on vacation, at the clinic or in the hospital. Be sure to ask everyone around you, including health care workers, to wash their hands, too!

When to Clean Your Hands

Whenever your hands look dirty, use soap and water, not just hand gel.

You should clean your hands often, especially at the times listed here:

- After coughing or sneezing or blowing your nose
- Before eating
- Before and after preparing food
- Before giving and taking medicine
- Before and after breathing treatments

Cleaning with Soap and Water

1. Wet hands with warm water.
2. Apply soap. Liquid, antibacterial soap is best.
3. Rub hands together to lather.
4. Scrub hands well,back and front, up to wrist and between fingers. Clean under nails. This should take 20 seconds to complete. (This is about the time it takes to hum or sing the "Happy Birthday" song from beginning to end, twice.)
5. Rinse in warm, running water.
6. Use a clean paper towel to dry your hands completely.
7. Turn off the faucet by using the paper towel.
8. Throw paper towel away.



Cleaning with Hand Gel

1. Be sure the hand sanitizer is alcohol-based (at least 60 percent alcohol).
2. Apply the sanitizer to the palm of one hand.
3. Rub hands together.
4. Rub the sanitizer over all surfaces of hands and fingers.
5. Keep rubbing until hands are dry.
6. Rubbing should take about 15 seconds.
7. Remember, if you see that your hands are dirty, wash with soap and water.

- Before and after airway clearance
- Before touching your face or eyes or putting in contact lenses
- After being around someone who is sick
- After going to the bathroom
- After changing diapers or cleaning up a child who has used the toilet
- Before and after clinic visits
- After touching surfaces in the hospital room
- After touching garbage
- After touching an animal or cleaning up after your pet

Cough Hygiene — Cover Your Cough!

Many serious respiratory illnesses, including the flu, are spread by coughing or sneezing. To help stop the spread of germs, cover your cough!

Cover your mouth and nose with a tissue when you cough or sneeze.

Put your used tissue in the waste basket.

If you don't have a tissue, cough or sneeze into your upper sleeve or elbow, not your hands.

Remember to clean your hands by washing them with soap and warm water or using an alcohol-based hand rub.

Cleaning Respiratory Equipment

Germs can get on respiratory equipment and cause lung infections. Clean and disinfect all equipment used for inhaled medicines or airway clearance by using the guidelines described here and in the manufacturer's instructions. It is vital to clean and disinfect equipment such as nebulizers that have been in contact with mucous membranes, sputum or phlegm. To clean and disinfect your nebulizer, follow these steps:

1. Clean your hands.
2. Clean the nebulizer parts.

   The nebulizer parts must be cleaned before they can be disinfected. With a new paper towel, wash the inside and outside of the nebulizer parts with clear liquid dish soap and hot water. Be careful not to damage any of the parts.

   Throw the paper towel away, then rinse the nebulizer parts with water. Clean the nebulizer right after it is used to keep the medicine and ***debris*** from drying. Once debris dries,

it is difficult to wash off. You also can clean the nebulizer parts in an automatic dishwasher if the nebulizer's manufacturer's instructions allow.

3. Disinfect the nebulizer parts.

   DO NOT USE VINEGAR. Vinegar is not strong enough to kill the germs a person with CF might get.

   Instead, ask your CF care team what is the best way to disinfect the nebulizer parts. Some options are:

- Using an electronic steam sterilizer (e.g., used for baby bottles)
- Boiling (in water) for 5 minutes
- Microwaving (in water) for 5 minutes
- Washing in dishwasher, if the water is hotter than 158° F, for 30 minutes
- Soaking in 70 percent isopropyl alcohol for 5 minutes
- Soaking in 3 percent hydrogen peroxide for 30 minutes

   Read the manufacturer's instructions to learn about cleaning and disinfecting your nebulizer. Do not use a nebulizer that cannot be disinfected.

4. Rinse the nebulizer parts.

   If you disinfect with isopropyl alcohol or hydrogen peroxide, rinse all parts well with sterile water.

   DO NOT USE WATER FROM THE FAUCET, BOTTLED WATER OR DISTILLED WATER. You can make water sterile by boiling it for 5 minutes. Use this water once, then throw it out. If you disinfect by other methods, you do not need to rinse the nebulizer.

5. Air-dry the nebulizer parts.

   After the final rinse, drain the parts on a clean surface covered with new paper towels. Replace wet paper towels with dry ones and fully air-dry all parts. Germs will grow on anything that stays wet. Store the dry nebulizer in a clean, dry bag in a clean, dry place.

Some respiratory equipment may need to be cleaned but not disinfected. These items can be cleaned often with liquid soap and hot water. Ask the respiratory therapist, the nurse or physician at your CF care center, how often to clean your equipment and the best way to do so.

When People With CF Live Together



People with CF who live together should do airway clearance in separate rooms and not share respiratory equipment.

People with CF who live together can get germs from each other. To decrease the spread of germs, they should limit contact with each other's mucous membranes, sputum or phlegm.

To decrease the risk of getting germs from each other, people with CF should **NOT** share:

- Respiratory equipment
- Airway clearance devices
- Toothbrushes
- Eating utensils or drinking cups
- Anything that has been in contact with mucous membranes, sputum or phlegm

When a person coughs, germs can spread 6 feet or more. Doing airway clearance at different times and in different rooms can help decrease the spread of germs.

Being With Other People With CF

People with CF may benefit from knowing others with CF. These benefits include support, friendship, advice and having a positive role model. Many also benefit from feeling that they are not alone in their fight against CF.

However, people with CF can pass germs to each other. Some germs are worse than others. There are many different types of germs out there. For example, *Pseudomonas* and *B. cepacia* complex are both types of bacteria that can be passed between people with CF.

People with CF who do not live together should avoid face-to-face activities and those that spread germs, such as hand-shaking, hugging or kissing. Keeping your hands clean helps prevent the spread of germs.

At School

If your child has CF, consider letting the school nurse know. Work with your CF care team to decide what information to tell the school. Members of the CF care team can help you talk to the school about CF. More than one child with CF may attend the same school. They should not be in the same classroom. To lower exposure to germs, ask the school to schedule common activities, such as lunch, at different times. Again, hand hygiene can help prevent the spread of germs that cause infections.

Camps

CF germs have been passed between people with CF at CF-specific camps. People with CF should not attend CF-specific camps. The risks of getting CF germs outweigh the benefits. Because it is vital to exercise, make friends and build support systems, people with CF are encouraged to join in camps, social groups and sports activities that are not specifically for people with CF.

CF Foundation's Infection Prevention and Control Policy

The CF Foundation's Infection Prevention and Control Policy is focused on protecting and maintaining the health of people with CF at all Foundation events, meetings and offices.

Medical evidence shows that all people with CF could have germs in their lungs and sinuses that might be spread to others with CF. The infection prevention and control policy includes practices and activities to promote safety and reduce the risk of cross-infection among people with CF.

Besides cleaning your hands and covering your cough, the key elements are:

- At any CF Foundation-sponsored indoor event, meeting or office only one person with CF may be present. This person will be designated by the Foundation.
- At CF Foundation-sponsored outdoor events or gatherings, people with CF need to maintain a distance of at least 6 feet from each other.
- Under no circumstances shall individuals with CF who have ever had a confirmed positive sputum culture for *Burkholderia cepacia* (*B. cepacia*) complex attend any CF Foundation events, meetings or offices.

You can read the full policy on the CF Foundation's website: **www.cff.org**.

Germs in Public

Everyone should avoid unnecessary contact with people who have a cold, the flu or appear ill. This does not mean that children with CF should be kept from attending school or activities because parents are afraid the child will get sick or "catch" something. Nor does it mean that people with CF cannot go to college, have jobs and enjoy active full and lives.

Along with doing good hand hygiene and getting vaccinations, parents of children who have CF can help their children avoid germs by:

- Keeping children from play times or a sleepover with a child who is sick.
- Cleaning toys passed from another child to yours. Soak toys for 10 minutes in a solution of one quarter cup chlorine bleach and one gallon of water and rinse in plain water. Soft toys should be put through the washing machine regularly.
- If children are in day care, find out what the day care center's policies are on children with illnesses who attend. Ask how often the toys are cleaned.
- Beware of indoor play areas at malls, theme parks and other places where a lot of children pass through, such as a ball pit. The cleaning may be less than ideal.

People with CF need to:

- Avoid touching their eyes, nose and mouth, because germs spread this way.
- Stay away from others if you are ill, and stay away from people who are ill. This helps prevent the spread of germs.
- Avoid spas and hot tubs. *Pseudomonas* has been found in spas and hot tubs.
- Avoid pools without enough chlorine to kill *Pseudomonas*. Your city or county public health department can tell you what chlorine levels kill germs and meet public health standards.
- Consider your work environment:
  - Work involving toddlers and school-age children — for example, work in day care centers and elementary schools — increases your exposure to respiratory viruses and may increase the frequency of CF lung infections.
  - Work in a health care setting may expose you to germs that are potentially harmful to your lungs.

Infection Prevention and Control in the CF Care Center Clinic and Hospital

"Infection control" or "infection prevention and control" is a term used in the hospital or clinic setting. It refers to the policies and procedures used to minimize the risk of spreading infections. All CF Foundation–accredited care centers have infection control policies and procedures in place to protect people with CF.

In addition, guidelines called "standard precautions" are recommended by the CDC for reducing the risk of spreading of germs from blood and other sources. Standard precautions apply to all people receiving care in the clinic or hospital regardless of their diagnosis or whether they have an infection. Standard precautions apply to blood and all body fluids except sweat. This means that health care workers should use gloves and gowns and sometimes eyewear when they come into contact with blood and body fluids.

You can read the guidelines for infection control on the CF Foundation's website: **www.cff.org**

Talk to your CF care team if you have any questions or concerns about your CF clinic's infection control policies.

Remember

Knowing how to avoid germs can help people with CF stay healthier. Everyone should get all recommended vaccinations; this includes the flu shot. All people with CF should avoid unnecessary contact with people who have an illness, such as a cold or the flu. Good hand hygiene helps prevent the spread of germs. But no one can prevent exposure to all germs. Do not panic when you or your child gets a cold. You cannot avoid all germs without avoiding life. Children and adults with CF should live life to the fullest. Mental and emotional health depends on a normal, healthy contact with the world.

The following resources have helpful information about avoiding germs:

- The CF Foundation (**www.cff.org**):
  What You Should Know About Germs
  Germs and Infection Control (webcasts)
- The Centers for Disease Control and Prevention (**www.cdc.gov**):
  Handwashing: **www.cdc.gov/handwashing**
  Vaccines: **www.cdc.gov/vaccines**

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **All people with CF who are 6 months and older and who are not allergic to eggs should get the influenza vaccine every year before flu season starts.**
   a. True
   b. False

2. **Which of the following statements are true?**
   a. Children with CF should not be in group day care.
   b. Hand hygiene is an important part of preventing illness.
   c. People with CF cannot pass germs to others with CF.
   d. Alcohol-based hand gels is one way to clean hands.

Answers

To learn more, turn to the page number shown after the answer.

1. A

It is **true**. ALL people with CF, who are 6 months and older and are not allergic to eggs should get the flu shot every year before flu season starts. In fact, household members, relatives and close friends of people with CF; day care staff and health care staff should get the flu shot as well! Page 103.

2. B, D

It is true that **hand hygiene is an important part of preventing illness and alcohol-based hand gels is one way to clean hands**. Children should live a normal life. They do not need to stay home for fear of getting sick. People with CF can pass germs to each other. Pages 105-106.

Notes and Questions

Chapter 10: Raising a Child with CF

Like any chronic condition, cystic fibrosis (CF) can cause social, emotional and psychological challenges. Understanding these issues can help you deal with them and raise a happy, healthy child.

Emotional Impact of CF

The diagnosis of CF can cause many emotions. This is normal. It is important to recognize them so you can work through them. Talking about your feelings with your family, friends and CF care team can help. It also may help to talk with a counselor or psychologist. Ask your CF social worker about resources and counseling options.

Some of the emotions you may feel include:

- Concern — for your child's health
- Worry — about the future
- Guilt — for having a child who inherited the disease from you
- Fear — of the unknown (How will I cope? How sick will my child be? How long will my child live?)
- Anger — that your child has CF
- Resentment — for the time and attention that the child with CF needs



The diagnosis of CF leads to a whole host of feelings. Understanding the impact of these feelings is the first step in learning how to deal with them and raising a happy, healthy child.

Keeping Life as Normal as Possible

Most people with CF live a normal daily life, with the challenge of incorporating daily medications, airway clearance techniques and other treatments and medications. Children with CF grow up, go to school, have friends, have hobbies and can exercise and play sports. Many go to college. Many marry and have families. The people with CF who do best are often those who were raised with the same expectations and rules as other children.

In guiding your child's emotional and psychosocial development, here are some basic rules:

- Treat your child as a normal child who happens to have CF.
- Do not be overprotective or neglectful.
- Foster your child's independence as he or she grows up.

All parents want to protect their children. It is vital for children to go through normal life events if they are to grow up and be well adjusted. Mental and physical health are closely related. Try to avoid being overly protective.



Let the child learn about nature, science, art, other cultures and the world.

One way to encourage independence is to help children learn about nature, science, art, other cultures and the world. Such children are less apt to be self-centered and self-pitying. If they can see that they are a part of a world that is bigger than their family and community, they will see that the world does not revolve around them. They also will know the joy of living and learning.

Foster a love of books by reading to your child. Reading takes a child to other places, far from the world of CF. It also is something to do during airway clearance and breathing treatments.

Teach your child empathy for others and for their challenges. Encourage your child to help others. Help your child see and appreciate all that is good in life.

Parenting at Major Transitions

Certain issues can be expected as your child grows:

Age 0–3: Playing in playgroups or play centers, and going to babysitters and/or day care centers are common at this age. Besides your child's normal developmental tasks, there are things to teach other parents, babysitters and day care staff about CF. This may include checking the cleanliness of the facility or showing the staff how to give enzymes or other medicines to your child.

Age 3–5: Children at this age are entering preschool and being cared for by babysitters. Consider a program where the child attends school all day for a few days a week so they can learn about eating and taking enzymes away from home and family.

Problems with mealtime behavior are common in preschool and school-age children. In children with CF, these may include refusing or avoiding food, disruptive behavior and refusing enzymes. Many parents feel pressure to get their child to eat.

Scolding, pleading, lecturing and forcing do not work and may make things worse. Your CF dietitian can give you some tips on how to have positive mealtimes and make good food choices. Good nutrition is a team effort with the family and CF care team. Let them know when you are stressed or need help with feedings or mealtimes.

Age 5–6: At this age, children enter kindergarten. With teachers, as with doctors, the goal is teamwork. Explain that you are glad to answer questions or offer help, but you want your child to be as independent as possible in taking enzymes before lunch and snacks. Ask the teacher to help you promote independence and self-care. Ask the teacher for suggestions. Mention that rarely are there classroom emergencies with CF.

Some of the issues to discuss with your child's teachers each year are:

- Coughing
- Frequent bathroom visits
- More water fountain visits (or the ability to have a water bottle)
- Taking enzymes at the beginning of all meals and snacks containing protein or fat
- Having extra snacks and extra portions of meals
- Medicines
- Absences when ill

It may help to send a letter to the teacher from you or the CF doctor. Ask your CF care team for information to give to the teacher. The CF Foundation has *A Teacher's Guide to CF* and a letter that may be helpful. For a free copy, go to **www.cff.org** or call **1-800-FIGHT CF**.

School-age children can often take their own enzymes and medicines. Parents should become comfortable with other people supervising their child taking enzymes.

Plan how to handle schooling and homework when a child is hospitalized. Many children with CF are in the hospital for a week or two, each year or so, for treatment of an exacerbation. Plan ahead so it does not interfere too much with school. Talk to the principal and teachers about how to handle schoolwork during absences and hospital stays.

Call your CF social worker for help in planning. There may be services such as tutoring or in-hospital schools that can help your child stay on top of schoolwork. Consider giving the school a written copy of this plan. The best way to help your child is to stay involved.

Sometimes, students with CF and their families choose to prepare an ***Individualized Education Plan (IEP)***, under the ***Individuals with Disabilities Education Act***, or a 504 Plan, under ***Section 504 of the Rehabilitation Act*** of 1973. These laws help people with CF get services they need to secure a free and appropriate public education while maintaining their health.

For example, IEPs can help establish ways that students can get special tutoring if they are absent because of illness or hospitalization. Contact your child's school at the start of each school year about getting an IEP. Your CF care center has a sample letter that your CF doctor can use to write to the school to explain how CF affects your child and to identify some things that may help.

To learn more about CF and school issues, talk to your care center team. The CF Foundation pamphlet "School and CF" is also available by visiting **www.cff.org** or by calling **1-800-FIGHT CF**.

How to Talk to Your Child About CF



Telling your child that he or she has CF may seem hard. Yet, understanding CF and why the treatments and medicines are so important helps your child prepare for the future. Your CF care team can help you explain CF to your child.

A child's first questions about CF often come after starting school. He or she sees that other children do not take enzymes or do daily airway clearance. They may be asked questions about CF by other children, adults or teachers.

The goal of telling your child about CF is to help your child know what CF means and why eating right, taking enzymes and other medicines, and doing airway clearance is important. Another goal is to get your child ready to do his or her own care.

The hardest question for everyone is about death. Other children may have heard that CF is a fatal disease of childhood, which it once was. One mother explained it this way: "We told him, 'There are a lot of ways a person can die. We do not know if you will die from CF or not. Right now, you have medicines that help keep you well. If you take your medicines and do what you are supposed to do, you will probably be okay, but CF is something you will have for the rest of your life.'"

Children should be taught about CF as soon as they can understand. What you say to your child depends on his or her age, personality and ability to understand. If you have questions, ask your CF care team for help.

Parents should teach their children how to talk about CF. The goal is for the child to be able to talk about it in a matter-of-fact way. For example, "I take these enzymes to help my body digest food"; "I do airway clearance to clear my lungs"; or, "The reason I cough is to help my lungs." Your CF care team can help you teach your child about CF and how to answer questions.

What About Discipline?

Many parents of children with CF are concerned about discipline. Many are hesitant or feel guilty about disciplining their child. Discipline is vital for any child to be well adjusted and psychologically healthy.

It is important that you apply the same discipline and standards to all children in the family. If you have other children who do not have CF, they may resent what they see as special treatment for the child with CF.

It helps to teach and model social manners. You want to avoid having a child with a chronic illness who acts "bratty." Good manners help any child to be accepted.

Sometimes, your child may refuse treatments. Explain how important the treatments are for his or her health. Make the treatments pleasant and fun by playing music, playing games, reading or watching TV during treatments. The best way for your child to learn how vital treatments are is for you to be consistent in giving them. If they still refuse, you may need to withhold "privileges" such as TV or video games to motivate cooperation.

If you need help, talk with your child's CF doctor, social worker or psychologist about a referral to a behavior specialist. Do not give up: It is important for your child's health!

Siblings (Brothers and Sisters)

Although siblings of a child with CF may feel ignored or left out at times, some siblings also learn to care about others. Children with CF get extra attention because of the treatments. Teach siblings about CF to help them understand why the child with CF may get extra attention. Books about CF, like this one, are good for siblings and relatives. To help siblings feel important, spend some time with each one alone every day. Allowing the siblings to attend clinic visits or help with treatments will also help them feel involved in caring for the child with CF.



Siblings of a child with CF may feel left out because of the extra attention being given to the child with CF. Your CF care team can help you explain CF to all family members.

School Friends and School Issues

When children start school, they make new friends and develop a social life. Now you are dealing with classmates, playmates and their families. It is hard to decide how much to tell others about CF. Most families find a matter-of-fact approach is best. School friends and their families may need to know that CF is not contagious and that your child's cough is not a symptom of something that their child can get.



Classmates and friends join a child with CF at a CF Foundation Great Strides event. When questions are asked about CF, encourage your child to give a brief explanation and let people know that they cannot get CF from someone else.

If sleepovers, long visits or meals together are planned, make sure your child's enzymes, other medicines and treatments are in the plans. However, once in a while your child might miss an airway clearance treatment or a dose of medicine so that he or she can be involved in activities with a friend. It may be better for your child's psychological and social well-being to go to a friend's house than to stay home and not miss a treatment. But make sure your child does not skip treatments routinely.

It is important to speak to your child's teacher about CF and about the importance of infection control for your child's health.

Please keep in mind that the germs often carried by children who do NOT have CF can be harmful to those with CF. Therefore, a child with a cold or flu should be encouraged to stay home. If a child with an illness is in the classroom, he or she should do hand hygiene frequently and use tissues when sneezing, coughing or blowing his or her nose.

CF-specific germs, such as *Pseudomonas aeruginosa* and *Burkholderia cepacia* complex, usually are not harmful to people who do not have CF. Therefore, a child with CF who has a lung infection with these germs cannot pass it on to a child who does not have CF. However, these germs can be harmful to others with CF. If there is more than one person with CF in the school, such as another student or a teacher, special attention to infection control guidelines should be followed. (See Chapter 9 for more information about avoiding germs.)

How to Handle Unwanted Questions

Just because your child has CF does not mean you have to open your life up to everyone. Teach others about CF, but remember that you and your family have a right to privacy. You may have to deal with unwanted questions from concerned or nosy people you meet in public places. They may ask questions or make comments about your child's cough. There are many ways to deal with unwanted questions or comments:

- Ignore them.
- Smile and shrug.
- Say, "My child has a lung problem that causes coughing."
- Say that your child has CF and explain a little about it.

Building a Relationship Between Your Child and the CF Doctor

Doctor and clinic visits can be scary for young children. Reading children's books about doctor visits can help, as can a toy doctor's bag for play doctor visits. As your child gets older, go over their questions before each doctor visit. Encourage your child to ask questions. Role play to practice the visit. During the visit, try to keep your child involved in the conversation. Let the child answer the doctor's questions. You can add the details later. This helps teach children independence in their care.



It is important to help your child build a good relationship with the CF doctor.

Hospital Stays

When a child is in the hospital, parents should talk to the staff about their level of involvement in their child's care. Some care has to be done by the staff; some can be done by parents or other family members. Talk to the CF care team and hospital staff about what is best for your child. Remember, you are part of the team working to help your child get better.

During hospital stays, bring things from home to make you and your child more comfortable (toys, videos, favorite stuffed animal, pillow, favorite blanket or books). Check with the nurse to see what you can bring and to get other ideas to make your child's hospital visit better.

Review Questions

Answers follow the questions. More than one answer may be correct.

1. **Who can help you find ways to work through the emotional impact of having a child diagnosed with CF?**
   a. Family
   b. Friends
   c. CF care team
   d. Counselor or psychologist
   e. All of the above
2. **Which of the following are true statements about raising a child with CF?**
   a. Treat your child as a normal child who happens to have CF.
   b. Teach your child about CF as he or she grows.
   c. During a hospital stay, parents are a part of the team working to help the child.
   d. All of the above

Answers

To learn more, turn to the page number shown after the answer.

1. E

All of the above are true. Other people can help you work through the emotional impact of a CF diagnosis. The diagnosis can cause feelings of concern, worry, guilt, fear, anger and resentment. These feelings are normal. Page 115.

2. D

All of the above are true. Your child is a normal child who happens to have CF. Teach your child about CF as he or she grows. During a hospital stay, parents are a part of the team working to help the child. Pages 115, 118, 121.

Notes and Questions

Chapter 11: Adolescence and CF

Adolescence (the teen years) is the time of transition from being a child to being an adult. As you grow and your body changes, and you gain more freedom, you also take on more and more responsibility.This means that many of the CF-related tasks that parents once took care of will become the teen's job. The CF care center team is there to help with this transition.

Staying Healthy

Dating, learning how to drive, working your first job and choosing a path for higher education or job training beyond high school are just a few of the exciting milestones of adolescence. You should also plan to reach for your goals. It is more important than ever that you stick to your CF treatment plans.



Doing CF treatments every day will help keep you healthy and help you reach your goals.

You want to enjoy your life now and have fun with your friends, and it may seem as if CF treatments are getting in the way of your fun. But, remember, if you get sick, you will have even less time to do fun things. Staying healthy and avoiding CF complications by sticking to your treatments will help you enjoy life more. If you feel stressed out about the amount of time needed each day for CF treatments, or if you are wondering why you need to do a particular treatment, talk about it with your CF doctor or nurse. Perhaps you can work together to find a way to make treatments less time-consuming.

Young people with CF go through puberty and develop into men and women just as other people without CF do. When puberty starts may depend on the overall health of a person. Teens with CF with good nutrition will usually have a normal growth spurt. Having enough body fat helps puberty start. Getting enough calories is very important to support all of the physical growth and changes you are going through.

No one should smoke — especially not people with CF. Smoking harms just about every part of your body, but it is devastating for your lungs, which already have enough to deal with from CF. Some adolescents with CF, if under peer pressure to smoke, find it works well to use their CF as an excuse to say no. You also need to stay away from secondhand smoke, or breathing in smoke from other people's burning tobacco. If you smoke, ask a member of your CF care center team to help you find ways to stop.

Most teenagers experience intense emotions at times, including sadness, anxiety and confusion. This may sometimes be even more true for teenagers who have with CF. If you are feeling overwhelmed by your emotions, talk to someone you trust, such as a family member, a friend, a teacher, your school counselor or a member of your CF care team. The social worker or psychologist at your CF care center can be helpful in times like these.

Sports and Hobbies

Most people with CF have normal experiences with school, sports and hobbies. The more exercise and activity you do, the better for your health. Exercise helps keep lungs healthy, improves your appetite, reduces stress and makes you feel good. Remember, when exercising or playing sports or when you are out in the heat, people with CF lose more salt in their sweat than others. You should carry water bottles when exercising or when in the heat, and stop for a big drink of water at least every half hour. You should also eat salty foods before and after exercise.



People with CF should do activities they enjoy.

Talk to your CF doctor or physical therapist before starting an exercise program about what type of exercise is appropriate for you, how long it is safe to exercise and whether there are any precautions. If participating in highly active sports is not possible for you, look for other activities and clubs you can join to share your interests and skills with your peers at school and in your community.

Your Identity and CF

Yes, you have CF. But CF does not define who you are. During adolescence, you are developing your identity — a continuing process of learning, exploring your interests, developing your strengths and interacting with others to figure out who you are. It is a very exciting time, but it also can be a time of great anxiety for any adolescent.

Adolescents with CF may have unique worries. They may be self-conscious about their cough. They may be smaller than their friends. They may tire more easily. They may have to take medicines or treatments at school. They may worry that no one will want to date them because they have CF.

Your discomfort may be eased by learning as much as you can about what CF is, the challenges it can bring and why the treatments and medicines are needed. It will help if you can explain to friends that:

- The cough is not contagious and helps you clear your lungs.
- Medicines help digest food, fight respiratory infection or provide vitamins.
- CF is what you have, not who you are.

Most adolescents find that being open about having CF makes life easier, especially with close friends and people they date. Letting friends know about CF can prevent some awkward moments, such as having to explain why you go to the bathroom more or need to take so many pills with meals. Many adolescents who have been open with friends say it does not change their friendships. Removing the mystery helps build acceptance and understanding. Role playing questions and answers with parents or siblings may also be helpful.

Becoming Independent

Adolescents and young adults want independence. They may rebel against schedules, treatments, medicines and limits. They may refuse to do regular airway clearance. It is normal for you to want independence. Learning independence and responsibility is a vital part of growing up. However, refusing to do your CF therapies can cause serious — even life-threatening — harm to your health.

Work with your parents and the CF care team to gradually shift the bulk of responsibility for CF care from your parents to yourself. Some of the things you must start to do include:

- Learn about your treatments, including the names of your medications, what dosage to take, what time to take and how often to take.
- Learn how to take care of your airway clearance equipment, including cleaning it correctly.
- Call the CF clinic yourself to ask questions.
- Write down questions for your CF care team before your clinic appointments.

- See the doctor first alone during CF care center visits, then with your parent or guardian.
- Get to know the ins and outs of your health insurance plan.

Transferring to the Adult CF Clinic

Toward the later part of adolescence, most people with CF transfer from a CF clinic that specializes in treating children to one that specializes in treating adults with CF. It may be a little sad and scary to leave the clinic and the people who you know so well, but adults with CF are different from children with CF. The staff at the adult CF clinic are experts in caring for the special health care needs of adults with CF. The transition should be a planned process over time, not an abrupt change.



Planning for the Future

Adolescence is the time to start planning for the future. Even though you have CF, you should be thinking about education and jobs just like your friends without CF are. Start thinking about your future education or job training at the start of high school, not at graduation. There are scholarships available specifically for people with CF. There also are programs to help people with CF cope with the stress of high school and college or to find job training. Your CF social worker can help you find these programs.

Colleges, universities, vocational training centers and other places of higher education that get federal funds cannot refuse to admit a qualified student solely because the student has a disability such as CF. Most colleges and universities have an office for students with disabilities that can help students get the accommodations they need. You do not have to tell the school that you have CF unless you request accommodations because of CF.

After you graduate from higher education, make sure that the employer you choose offers a health insurance plan and other benefits, such as sick leave, that fit your needs. The social worker at your CF care center can help you with this. You can also find more information about what to look for in a health insurance plan on the CF Foundation website, **www.cff.org**.

For more information about school and adulthood with CF, see the resource list in Appendix B, page 183.

Remember

The teen years are a time of incredible growth for your body and mind. You will take on more and more responsibility for CF care and other tasks as you get closer to adulthood. Keeping yourself healthy by sticking to your CF treatments will help you make the most of this exciting time and prepare for the future. Base your developing identity on your interests and strengths, not your CF. Talk to your CF care center team about how to find a healthy balance between CF care and the other parts of your life.

Notes and Questions

Chapter 12: Adult Life with CF

People with CF are living longer and healthier lives. As those with CF move into adulthood, new issues develop concerning independent living, marriage, family planning, career and finances. It can be hard to address these issues at the same time that a person is managing a chronic condition. People with CF should be encouraged to settle into an adult lifestyle, mixing good health care with independence, marriage, family and career. Your CF care team can help you with these issues.



People with CF are now living longer and healthier lives and they are facing new challenges.

Transition Issues

Most people with CF transition from pediatric to adult care between the ages of 17 and 21. This means changing from a pediatric to an adult CF care setting. This transition should not happen abruptly, but instead should be a planned process over time. During this transition, the responsibility for CF care moves from parents to the person with CF. Adult clinic staff usually talk with parents only after getting the adult with CF's permission. They also deal with adult issues, including marriage, parenthood and careers.

Independence

Most adults with CF live on their own and do their own CF care. Some airway clearance techniques offer more independence with treatments. Ask your CF respiratory therapist which technique is best for you.

During illness, adults with CF may need help with their care. The CF care team, family or friends may be able to arrange for help and support during these times.

Family Issues

Young adults with CF may deal with marriage and family issues. Even though CF does not affect a person's ability to participate in sexual activity, CF can make it harder to get pregnant. Women with CF may be less fertile than women without CF. Ninety-eight percent of men with CF are infertile. This can make family

planning stressful. Your CF care team can arrange a fertility evaluation so you can find out your fertility status and determine whether any medical help will be needed.

For a woman with CF to have a successful pregnancy, it is important that she has good lung function and nutritional status. If you are thinking about getting pregnant, talk to your CF doctor about what you need to do to make sure your health is strong enough. Also, if you get pregnant, it is important to have some help with child care in place before the baby is born so that you are able to also care for your own health and the baby.

Because a person with CF has two copies of the CF gene mutation, any biological child he or she produces will inherit one copy of the gene mutation. (It takes two copies for a person to have CF.) Therefore, if the other person in the couple does not have CF, and it is not known whether he or she carries a CF gene mutation, some couples may decide to have that person tested so that they know how likely it is that they will have a child with CF. However, the test cannot detect every CF gene mutation. Your CF care center can help arrange genetic testing and genetic counseling. Some couples decide that adoption is the best choice for them.

It is important to keep in mind that if people with CF do not wish to become pregnant, they still should use contraception. People with CF also need to use barrier protection, such as a condom, to protect against sexually transmitted diseases, just like anybody else would.

See Chapter 6 in this book for more detail about the reproductive system in people with CF.



People with CF should plan for the future like everyone else.

Career

Just like other adults, those with CF should look for a satisfying job or career that uses their special skills and interests. However, adults with CF must manage the effects of chronic illness on their careers. Treatment schedules and medicine routines must fit into work schedules. Doctor visits and illness may cause missed work days. Some adults with CF seek employers who offer flexibility or choose to be self-employed. Access to quality health insurance is another important consideration. The CF care team can help with treatment and work schedules and can help educate employers about CF.

People with CF should use common sense with regard to their work environment. For example, people with CF should select work that avoids exposure to lung irritants, such as smoke, dust and chemicals. Work with young children, such as in a day care center, and certain health care settings could expose a person with CF to germs that may cause respiratory infections.

Talk to your CF social worker and care team about what to look for and what to avoid in the workplace, as well as your legal rights, and whether or when to share your CF diagnosis with your employer. For more information about your legal rights, see the resources in Appendix B in the back of this book.

Remember

As they grow up, people with CF should plan ahead to have the same experiences as adults without CF, including living independently, a career, marriage and children. Choose your own unique path, and work with your CF care team to figure out what you need to do to fulfill your dreams in a way that also maintains your health.

Notes and Questions

Chapter 13: The CF Care Team: Who They Are and What They Do

People with cystic fibrosis (CF) need a team approach for all aspects of their care. At Cystic Fibrosis Foundation–accredited care centers, the team includes the person with CF, the family and several members from different medical disciplines and specialties. With so many people on the CF care team, it is easy to get confused. This chapter describes the members of the CF care team.

Who Is on the CF Care Team?

The most important members of the CF care team are the person with CF and his or her family members. The clinic part of the team is headed by the physician who sometimes is the CF care center director. The CF Foundation requires that the following medical people are part of every CF Foundation–accredited care center:

- Physician/CF care center director
- Program/clinic coordinator
- Nurse
- Dietitian
- Social worker
- Respiratory or physical therapist

Many CF care centers also have other specialists on the care team. These members may include:

- Other physician specialists
- Doctors in training
- Nurse practitioners
- Physician assistants
- Clinical nurse specialists
- Pharmacists
- Psychologists
- Child life specialists
- Chaplains
- Genetic counselors



Since CF is a complex disease, it requires a team approach. The CF care team consists of the person with CF, the family and several health care professionals.

Role of the People on the CF Care Team

People with CF, their families and CF health care professionals make up the CF care team. Every member of the CF care team has a vital role in helping provide the best personal care for the person with CF.

The Person with CF and His or Her Family

People with CF and their families are full partners of the CF care team in managing this chronic condition. Information and communication must flow to and from the person with CF and family members and other team members in an open and trusting environment. Every person with CF and his or her family members are able to be involved in care at the level he or she desires. Care is respectful of the persons' and families' preferences, needs and values.

Physician and CF Care Center Director



The team approach to CF care may seem confusing at first, but it gives you access to experts in CF care.

The CF care center director is the doctor who heads up the CF care center and health care team. Not all doctors at a CF center are the center director, but they are experts in how to care for people with CF.

The CF doctor works with the person with CF and his or her family, and the medical care team members, to make a treatment plan. He or she also teaches other doctors, health care workers and the public about CF. They may also do CF research. Your CF doctor will communicate and work with your primary care provider to provide the best care.

Program/Clinic Coordinator

The program or clinic coordinator is a health care professional who coordinates the CF health care team. He or she works with the director to be sure that the CF program runs smoothly. Much of what the coordinator does is "behind the scenes." If you have a question about how the clinic runs, you may want to ask the coordinator.

Nurses

CF nurses specialize in CF care. CF nurses fill a variety of nursing roles in CF care centers. Nurses are the primary contact for people with CF and their families. CF nurses work in the CF clinic setting, provide bedside care for people needing hospitalization and provide phone contact. Nurses have the primary responsibility for providing CF education at initial diagnosis and throughout life.

CF nurses:

- Help coordinate and carry out health care plans.
- Aid communication between the CF care team and the person with CF and family.
- Alert team members to psychological, social and financial concerns.
- Educate the person with CF, the family, the public and other health care workers.
- Support the person with CF and his or her family.

Dietitians

A dietitian (RD) is trained to look at a person's diet, growth and overall nutrition. A dietitian can teach people with CF and their families how to adjust their diets so that the person with CF can get the calories and nutrients he or she needs to be healthy. Dietitians help people with CF and their families keep the person with CF well nourished. They advise about:

- Baby formulas
- High-calorie, high-protein diets for people of all ages
- Supplemental vitamins
- Pancreatic enzyme supplements
- Dietary supplements

Social Workers

Social workers (SWs) help people with CF and their families with the social, emotional and psychological impact of dealing with CF. The social worker helps families deal with situations and issues that may interfere with their ability to handle health problems. He or she aids people to figure out how to get help with health care insurance, career choices and school issues. The social worker helps people with CF and their families by:

- Providing assessments and counseling regarding the psychosocial aspects of CF.
- Aiding communication between the family and other members of the care team.
- Teaching ways to cope with the stress of a chronic condition.
- Providing help to people with CF and families as they negotiate multiple systems, including insurance and employment.
- Finding financial assistance for families.
- Preparing teens and young adults for independence.
- Identifying family stress that may need professional help.
- Supporting family-centered care to provide optimal CF care.

Respiratory Therapists

Respiratory therapists (RTs) plan, teach and carry out respiratory care programs. In the hospital, RTs care for people with CF, giving airway clearance and aerosol treatments. RTs teach people with CF and their families how to do these treatments at home. They are also responsible for the care and use of equipment, including nebulizers, air compressors and oxygen systems. The RT does pulmonary function tests (breathing tests).

Physical Therapists

Physical therapists (PTs) design and direct exercise and activity programs for people with CF. A PT is trained to help a person make an activity and 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.

Other Physician Specialists

Physician specialists are also referred to as subspecialists. Physician specialists have extensive training and practice in a particular field of medicine or surgery.

Pulmonologist

A ***pulmonologist*** is a doctor who has special training in the diagnosis and treatment of diseases of the lungs. Some specialize in the care of children and teens, and others specialize in the care of adults.

Gastroenterologist

A ***gastroenterologist*** is a doctor who has special training in the diagnosis and treatment of diseases of the digestive system. This includes problems with the esophagus, stomach, intestines and liver.

Endocrinologist

An endocrinologist is a doctor with special training in the diagnosis and treatment of diabetes and other hormonal diseases, including problems with thyroid hormone and growth hormone.

Health Care Professionals in Training

If the person with CF is cared for at a CF Foundation–accredited care center that has training programs, he or she may see other people who are learning about CF. These may include other doctors, medical students and health care personnel who are learning about CF and how best to treat people with CF.

Fellows

A fellow is a doctor who has finished medical school and residency and is getting specialty training. The primary or "attending" CF doctor supervises fellows. At CF care centers, they are often pediatricians or internal medicine doctors who are specializing in CF and lung diseases. After years of special training, the fellow often goes to another CF care center to be a CF doctor.

Residents

A resident is a doctor who has finished medical school and received a medical degree. He or she is getting more training, often in pediatrics or internal medicine.

Medical Students

A medical student is working to become a doctor. Medical students are learning about diagnosing illnesses, caring for people and evaluating and reporting a person's condition and progress to the attending physician.

Nurse Practitioners

A nurse practitioner (NP) is a nurse who has an advanced degree with more education than other nurses. An NP is trained and licensed to do physical examinations and to prescribe medications and treatments. A pediatric nurse practitioner (PNP) is an NP who cares for children. An FNP is a family nurse practitioner. An ANP is an adult nurse practitioner.

NPs work closely with the CF doctor to plan and carry out a health care plan for each person and family. They also help the other nurses coordinate care for and provide education for people with CF and their families.

Physician Assistants

A physician assistant (PA) is trained and licensed to practice medicine under the supervision of a physician. A PA can do physical examinations and prescribe medications. PAs work closely with the CF doctor to plan and carry out a health care plan for each person and family.

Clinical Nurse Specialists

Clinical nurse specialists (CNSs) are nurses who have an advanced degree with more education than other nurses. CNSs have special training with groups of people with special health care needs, often people with chronic conditions. CNSs work with other CF team members to coordinate and manage care and provide education for people with CF and their families.



Pharmacists

A pharmacist helps manage the use of medicines in the hospital and home. A pharmacist looks to make sure medicines being prescribed do not react with each other and checks the dosages to help avoid errors. A pharmacist can show people how to take medicines and tell them what possible side effects to watch for.

Psychologists

A psychologist is a health care professional with expertise in assessing and treating problems with behavior, learning, emotions and group/family interactions. A psychologist helps with such problems as depression, learning disabilities or behavior issues.

A CF team psychologist may also focus on the problems people might 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. Psychology services may be brief consultations during regular CF clinic visits or during CF hospitalizations, or may require referral for ongoing psychology services. Psychologists help families and people with CF by providing:

- Adjustment to diagnosis (directed toward parents of newborns, and children and adults who are diagnosed later)
- Behavior management with young children (both general and CF-related issues)
- Assessment of cognitive, academic and emotional functioning
- Assistance with issues related to emotions, stress and being able to stick to the CF treatment plan

Genetic Counselors

A genetic counselor is a medical professional trained in genetics. A discussion with a genetic counselor can help a person understand inherited conditions and how an inherited disease is passed on to one's children. For example, families with a history of CF may learn their risk of having a child with CF through a genetic counselor.

Child Life Specialists

Child life specialists are trained to help children and their families cope with medical conditions and the tests and treatments required in the clinic and 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 and treatments. Child life specialists:

- Plan activities and entertainment during clinic visits and hospitalizations to provide ways to distract children from the stress of the medical treatments and the clinic or hospital environment.
- Teach skills for dealing with the stress of a chronic illness.
- Teach and direct medical play therapy to help children understand treatments and procedures.
- Provide a space in the clinic and hospital that is free from medical procedures, to keep a space for children that is "normal" and "safe."
- Work with other team members to teach children about CF, its effects and its treatment.

Chaplains

Many CF care centers have chaplains on the CF care team. They assist the person with CF and his or her family with spiritual issues.

Remember

The multidisciplinary team approach to CF health care may be confusing at first because so many people are involved. But, remember, everyone is working together with you to provide the best care for you or your child. And the best way the team can succeed is through open communication with you — the person with CF and/or the family members. If you are not sure who is the correct person to answer a particular question, start with your CF nurse, who can direct you to the right team member.

Notes and Questions

Chapter 14: The Cystic Fibrosis Foundation

The Cystic Fibrosis Foundation, a nonprofit, donor-supported organization, is the world's leader in the search for a cure for cystic fibrosis (CF). The CF Foundation's mission is to find a cure for CF and to improve the quality of life for people living with the disease. It accomplishes this by working to provide access to specialized medical care and effective treatments for people with CF and by supporting research to help develop new CF drugs and therapies.

The CF Foundation also works on behalf of people with CF through its public policy and advocacy program and provides educational resources to people with CF and their families, the public and medical professionals to increase knowledge of CF.



CF Foundation–supported researchers are developing new therapies to treat CF.

Medical Care

The CF Foundation funds and accredits more than 110 CF care centers, with affiliate programs and child and adult CF care programs throughout the country. At these clinics, people with CF have access to state-of-the-art CF care delivered by a team of CF experts from multiple specialties. This approach helps ensure that people with CF have the best health possible. Each center is thoroughly reviewed by the CF Foundation's Center Committee before it receives accreditation and funding. The review for accreditation occurs every year.

More than 40 years ago, the CF Foundation started collecting patient data, such as height, weight and gender, to track health trends and find better treatments. More than 27,000 people with CF get care at CF care centers across the United States. Patient data are kept in the confidential Patient Registry database. The registry provides the opportunity for wider insight into the disease and helps identify the best treatment methods and improve the quality of care.

Every year the CF Foundation publishes a Patient Registry Annual Data Report, which provides information on the health of people with CF and how much it has improved over time. In 2001, the Patient Registry was used to look at the overall health of people with CF by care center. The information showed that there were differences between care centers. Because of this, the CF Foundation began its quality improvement (QI) work. This effort is focused on speeding up the rate of improvement in care throughout the CF care center network. The work includes training care center staff in QI and helping make the partnership between people with CF, families and care center staff stronger.

To improve CF care, the CF Foundation and care centers work to reach the following goals:

1. People with CF and their families will be full members of the care team.
2. People with CF will have normal growth and nutrition.
3. People with CF will get the right therapies to keep lungs healthy and have fewer respiratory infections.
4. People with CF, their families and CF health care professionals will work together to decrease the spread of germs among people with CF.
5. People with CF will be followed closely to find and treat complications early.
6. People with CF and their families are supported when facing decisions about transplantation and end-of-life care.
7. People with CF will get care no matter their race, age, education or insurance coverage.

An important part of the QI work is to create up-to-date CF care guidelines to be used in all CF care centers. Care or practice guidelines recommend treatment based on published reports of clinical trials that look at safety, effectiveness and potential benefit. The guidelines include CF-related topics about nutrition/gastrointestinal health, respiratory health, infection control and more. The CF Foundation brings together experts on specific subjects, including doctors, nurses, respiratory and physical therapists, dietitians, adults with CF and parents of a child with CF to write guidelines on each topic.

The CF Foundation also has a Quality Improvement Toolkit. This toolkit helps people with CF, their families and care centers become stronger partners in improving the quality of CF care. For more information, visit the CF Foundation's website (**www.cff.org**.)

Patient Assistance Resource Center



The Cystic Fibrosis Patient Assistance Foundation (CFPAF), a subsidiary of the CF Foundation, is a nonprofit organization that helps patients and their families living in the United States afford medications and devices they need to manage CF.

For eligible enrollees, the CFPAF provides the following types of financial assistance:

- Payment of co-payments for approved prescription drugs and devices
- Payment of co-insurance for approved prescription drugs and devices
- Payment assistance with deductibles for approved prescription drugs and devices

The CFPAF also serves patients and families through:

- Insurance benefit verification and insurance counseling
- Reimbursement support for pre-authorizations and appeals
- Referrals to other patient assistance drug programs
- Referrals to alternative support programs, including rental assistance, local/regional assistance and other nonprofit organizations
- Patient advocacy and case management services
- An insurance access program to determine state and federal insurance eligibility, as well as Supplemental Security Income (SSI) and Social Security Disability Insurance (SSDI).

To learn more about the CFPAF, visit **www.CFPAF.org** or call **1-888-315-4154**.

Your care center social worker and the CF Foundation have more information available about private health insurance coverage and public assistance programs. To learn more, call **1-800-FIGHT CF** or **301-951-4422** or go to **www.cff.org** and look for the "Living with CF" section. More resources also can be found in Appendix B in the back of this book.

The CF Foundation also has the Patient Assistance Resources Library, an online database that provides up-to-date material to help people with CF, their families and CF health care providers work together on health insurance issues. Learn more at **www.cff.org/LivingWithCF/AssistanceResources/Library**.

The CF Legal Information Hotline provides free information about the laws that protect the rights of people with CF. Supported by the CF Foundation, the hotline is an important resource for people with CF who are facing challenges with health care coverage or reimbursement issues. You can contact the hotline at **1-800-622-0385** or **CFLegal@cff.org**.

For information about all of the Patient Assistance Resource Center programs offered by the CF Foundation, visit **www.cff.org/AssistanceResources**.

CF Services Pharmacy

The CF Foundation is affiliated with Cystic Fibrosis Services Inc., a Walgreens alliance partner. CF Services is the nation's leading provider of specialty CF drugs, case management services and reimbursement support for people with CF. For more information, visit **www.cfservicespharmacy.com**.

Research

The CF Foundation drives CF research forward. One way it does so is with its support of the CF research center network. These centers are located at universities and medical schools around the country. They bring together researchers who are experts in different areas of medicine. Scientists at these centers share ideas and information to increase what we know about CF. As knowledge about CF and how it affects the body has grown, researchers are finding new ways to treat the disease.

CF Drug Development Pipeline

The CF Foundation has a drug development pipeline of potential CF drugs that address all aspects of the disease. The CF Foundation started the Therapeutics Development Program in the late 1990s. This program gives scientists the financial support they need to participate in CF drug development and also creates opportunities for ***biotech*** companies to get involved in CF drug development.

Potential new drugs discovered through this work have shown encouraging results in laboratories and in clinical trials of people with CF. These results are very exciting, because some of these potential drugs aim to help correct the underlying cause of CF, instead of treating just the symptoms.

Clinical Trials

For CF treatments to be approved by the FDA, they have to be tested for safety and effectiveness in people with CF. The CF Foundation has an active network to help these studies be done more quickly. The Therapeutics Development Network (TDN) is made up of CF care centers that have training in doing clinical trials. The CF Foundation

has many resources to help people with CF learn more about volunteering for a clinical trial, helping develop new treatments and playing a role in the search for a cure.

To learn more about CF research, talk to your CF care center team, visit **www.cff.org/research** or call **1-800-FIGHT CF**.

Public Policy and Advocacy

The CF Foundation advocates on behalf of people with CF at all levels of government. It urges support for critical research, access to quality care and increased awareness of CF among state and federal lawmakers.

Bringing together a vast network across all 50 states, the CF Foundation connects people with CF, families, friends and other volunteers with their elected officials. The CF Foundation also supplies volunteers with the tools they need to advocate on behalf of people with CF.



Many people with CF and their families work directly with their elected officials on behalf of people with CF.

The policy and advocacy goals of the CF Foundation include:

- **Moving Research Forward**: Quickly move new treatments from research and development into the hands of people with CF by urging support for the national agencies engaged in drug development, such as the National Institutes of Health (NIH) and the FDA.
- **Improving Access**: Secure and protect the right of people with CF to benefit from high-quality private and public health care programs, such as Medicaid, and work to decrease the financial burden of the disease.
- **Increasing Awareness**: Educate elected officials about CF and the needs of the CF community. This educational outreach is done though the Congressional Cystic Fibrosis Caucus. You can learn more about the caucus at **www.cff.org/GetInvolved/Advocate/CFCaucus**.

To help achieve these goals, the CF Foundation:

- Connects CF advocates with elected officials in Washington, D.C., and their home states.
- Educates key members of Congress on issues important to the CF community.
- Supports an advocacy network of 90,000 people with CF and their families and friends to work with their elected officials on important issues.

- Helps CF care centers work with federal and state officials to support laws that benefit the CF community.
- Works to help people with CF and their families get and keep insurance coverage.

Visit **www.cff.org/GetInvolved/Advocate** or call **1-800-FIGHT CF** to learn more.

CF Education

The CF Foundation works to increase knowledge about CF for people with CF, their families, the public and medical professionals. The CF Foundation has information for people with CF, families and the general CF community, including an e-newsletter, *Connections*.

These resources are available on the CF Foundation website at **www.cff.org** and at CF Foundation-accredited CF care centers. The CF Foundation's educational webcasts bring the CF community together in a virtual "CF Education Day" forum to learn from the experts about living with CF and the latest in CF research. The webcasts are on the CF Foundation website for viewing at any time at **www.cff.org/LivingWithCF/Webcasts**.

The CF Foundation sponsors the annual North American Cystic Fibrosis Conference (NACFC), an international medical conference where scientists, researchers, doctors, nurses, dietitians, respiratory and physical therapists, social workers and other health care professionals gather to learn the latest in research and practice. The CF Foundation also supports training of health care professionals at CF care centers.

The CF Foundation also helps increase awareness and knowledge of CF through social media channels such as Facebook, Twitter and YouTube. Members of the CF Foundation's online community can connect with thousands of others who have CF or are related to someone with CF, and can also learn more about CF Foundation resources, programs and advocacy activities. Visit **www.cff.org** to learn more.

See Appendix B for a listing of many educational resources, as well as more information about the CF Foundation's medical care, research and public policy programs.

Local CF Foundation Chapters

The CF Foundation has about 75 chapters and branch offices around the country, staffed by professionals who help raise funds to support CF research and care. Each chapter has a group of volunteers who help make the fundraising events, such as golf tournaments, black-tie dinners and the annual national walk, Great Strides, so successful. Chapters also offer opportunities for volunteers to help spread awareness of CF and meet others in the CF community. Parent volunteers often make friends and find support with other parents who are dealing with CF.

Visit **www.cff.org** or call **1-800-FIGHT CF** to find a chapter near you.

Notes and Questions

Chapter 15: The Future and CF Research

The Cystic Fibrosis Foundation was formed in 1955 by a group of parents who had children with cystic fibrosis (CF). At that time, little was known about the disease and no effective treatments were available. Since then, the CF Foundation has raised hundreds of millions of dollars to support research to develop new CF therapies. This research has led to great progress in understanding and treating CF, which has made a huge difference in the lives of those living with CF.

Today, many people with CF are living into adulthood, and research to find a cure has never been more promising.



New treatments developed with the support of the CF Foundation have helped improve the health and quality of life of people with CF.

The CF Foundation and Research

The mission of the CF Foundation is to find a cure for CF and improve the quality of life for people living with the disease. Nearly every CF drug available today was made possible because of the CF Foundation's support and its ongoing work with researchers to find a cure for CF.

The discovery of the CF gene in 1989 — the single most important discovery in CF research — was the result of an international research collaboration. With the CF gene in hand, CF Foundation researchers for the first time could look at the root cause of CF in their research efforts to find a potential cure.

In the 1990s, the CF Foundation created the Therapeutics Development Network (TDN). The TDN was created to bridge the gap between what has been learned in the laboratory and the development of new therapies. The TDN is a nationwide network of nearly 80 CF clinical research centers. These centers specialize in conducting clinical trials to evaluate the safety and effectiveness of new CF therapies.

CF Drug Development Pipeline

The CF Foundation created a way to measure the progress of developing therapies, called a "drug development pipeline." To make it more likely that effective new therapies will become available to people with CF, the CF Foundation makes sure that several types of potential drugs are in development at the same time.

CF is a complex disease, and therapies must target problems in different areas, such as the airways and the digestive system. Different therapies also work on different issues, such as infection, inflammation and thick mucus.

Some therapies focus on fixing the cause of CF, and some therapies focus on better treatments for the problems CF causes. The following are several areas of CF research currently in the CF drug development pipeline.

CFTR Modulation

One way to fix the problems caused by the CF gene mutation is to focus on the gene's protein product, called the CF Transmembrane conductance Regulator (or CFTR). People without CF make healthy CFTR protein. This protein's job in certain cells is to make a tiny channel for salt and water to move in and out of the cells (See "How Do Altered Genes Cause CF?," page 16). When this channel is defective, the balance of salt and water is lost and the body makes thick, sticky mucus.

Researchers have been looking for and testing treatments that will help the CFTR protein work in people with CF. This is called CFTR "modulation" (meaning to adjust or change the CFTR protein). Restoring the health of the CFTR protein targets the underlying cause of CF and works at the most basic level of the body.

In 2012, the U.S. Food and Drug Administration (FDA) approved the CFTR modulator Kalydeco™ — the first drug to target the underlying cause of CF — in people ages 6 and older with a specific mutation of CF that is found in 4 percent of people with CF in the United States. Research is in progress to discover and develop additional drugs that will work in people with other mutations of the CF gene.

Restore Airway Surface Liquid

In CF, changes in the balance of salt causes mucus to be thick and sticky. Some research targets ways other than CFTR modulation to improve the movement of salt in and out of cells. This approach will help the mucus to become thinner and, therefore, cleared more easily.

Mucus Alteration

Mucus alteration studies evaluate drugs for their effectiveness in thinning and clearing thick mucus from the airways. For example, Pulmozyme® is an inhaled medication that thins and loosens mucus in the airways of people with CF. Approved in 1993, Pulmozyme was the first new drug specifically developed for CF in 30 years. Other methods to alter mucus are being studied.

Anti-inflammatory

Part of the damage to the lungs in people with CF is caused by inflammation. Anti-inflammatory drugs are being studied to help reduce inflammation in CF lungs, and thereby slow lung damage.

Anti-infective

Anti-infectives are used to fight or control infections. They can be used to fight chronic infections as well as exacerbations. Researchers are searching for and testing anti-infectives that are particularly useful against the types of bacteria that affect people with CF, such as *Pseudomonas aeruginosa*. Researchers have developed anti-infectives especially for people with CF that are inhaled directly into the lungs. This way, they go straight to the place they are needed to fight germs.

Transplantation

Lung transplants are serious and difficult procedures. But the growing success rate has made them a treatment option for some with severe lung disease. One area of research includes ways to improve how well people with CF do after receiving a lung transplant.

Nutrition

Good nutrition means better health for people with CF. Research focusing on nutrition looks at better ways for people with CF to absorb fats and proteins and, therefore, get more calories for growth and energy. Different enzymes and specially formulated nutritional supplements and vitamins are being researched.

For a current "snapshot" of therapies being studied, see **www.cff.org** and click on Drug Development Pipeline in the Quick Links or ask your CF care center.

Ask your CF doctor or research coordinator about a clinical trial you or your child might join.

CF Clinical Trials



A clinical trial is a health-related research study in human beings. Clinical trials follow a very specific study plan, which is called a "protocol." Protocols are carefully designed to protect the health of the person in the research study, while answering a specific research question. The research question is usually to find out if a new treatment for people with CF improves their health.

A protocol describes who can take part in the clinical trial; the schedule of tests, procedures, medications and dosages; and the length of the study. People who participate in the clinical trial are seen regularly by the research staff to monitor their health and to determine the safety and effectiveness of their treatment.

The clinical trial team includes doctors and nurses, and may include other health care professionals such as respiratory therapists. The research staff at CF care centers may be people

that you have already met who are on the CF care team. They may also be new to you if they work only in the research part of the center.

If you are thinking about joining a clinical trial, always think carefully about what the benefits and risks of participating in a specific clinical trial may be. Each specific clinical trial will have very specific risks and benefits.

In general, benefits for the person participating in the study may include:

- Playing an active role in one's own health care.
- Possibly gaining access to new research treatments before they are widely available. (Read about "placebos" later in this section.)
- Obtaining expert medical care at leading health care facilities during the trial.
- Helping others by contributing to medical research.

There are always possible risks involved in clinical trials that people must consider:

- There may be unpleasant, serious or even life-threatening side effects from the experimental treatment.
- The experimental treatment may not be effective for the participant.
- The protocol may require more of their time and attention than would a non-protocol treatment, including trips to the study site, more treatments, hospital stays or complex dosage requirements.

A placebo is an inactive pill, liquid or powder that has no medical effect. In CF clinical trials, the new experimental treatments are often compared with placebos. This is so the effectiveness of a new treatment can be compared with not receiving the treatment. During the study, the participant does not know whether he or she is receiving the new treatment or placebo.

Before participating in a clinical trial, people should know as much about the clinical trial as possible. The following questions might be helpful for the person thinking about being in a study to discuss with the health care team.

- What is the purpose of the study?
- Who is going to be in the study?
- Why do researchers believe the experimental treatment being tested may be effective?
- Has it been tested before?
- What kinds of tests and experimental treatments are involved?
- How do the possible risks, side effects and benefits in the study compare with those of my current treatment?

- How might this trial affect my daily life?
- How long will the trial last?
- Will hospitalization be required?
- Who will pay for the experimental treatment?
- Will I be reimbursed for other expenses?
- What type of long-term follow-up care is part of this study?
- How will I know that the experimental treatment is working?
- Will results of the trials be provided to me?
- Who will be in charge of my care?

There are many opportunities to help develop new drugs for CF. More potential therapies to treat CF are in development today than ever before. That gives all of us great cause for hope, but it also means more people with CF than ever before are needed to participate in CF clinical trials.

If you are interested in learning more about clinical trials, check with your CF care center. You may also learn more by going to **www.cff.org/research/ClinicalResearch/** and to **www.clinicaltrials.gov**.

Remember

As the understanding of the science of CF increases, there is also an exciting growth in the number of opportunities to discover and develop new potentially lifesaving therapies. Today, there are many trials underway studying an increasing number of potential new treatments for CF. With the CF Foundation's specialized network of CF research centers that have the expertise to do clinical trials, and the dedicated people with CF who want to participate in clinical trials, the future for people living with this disease has never been brighter.

Notes and Questions

PART IV

Resources



Appendix A: Medical Words to Know

A

Absorb/Absorption

The passage or uptake of substances into or across tissues, such as the uptake of digested food and water from the intestines into the bloodstream. Because of a lack of digestive enzymes in CF, some foods eaten by people with CF may not be well absorbed and used by the body. (Also see ***Digestive System, Enzyme, Intestine, Malabsorption, Pancreas.***)

Active Cycle of Breathing Technique (ACBT)

An airway clearance technique that involves using a set of different breathing techniques to loosen and clear mucus. (Also see ***Airway Clearance Techniques.***)

ACTs

See ***Airway Clearance Techniques***.

Aerosol

An inhaled medicine mist that treats lung problems. The aerosol or mist is created by a nebulizer attached to an air compressor. (Also see ***Air Compressor, Nebulizer.***)

Air Compressor

A machine that blows out a high flow of air through a tube. (Also see ***Aerosol, Nebulizer.***)

Airway Clearance Techniques (ACTs)

Different methods to loosen thick, sticky mucus in the lungs so it can be coughed or huffed out. (Also see ***Active Cycle of Breathing Technique, Autogenic Drainage, Chest Physical Therapy, Cough, High-Frequency Chest Wall Oscillation, Huffing, Intrapulmonary Percussive Ventilation, Oscillating Positive Expiratory Pressure, Percussion, Positive Expiratory Pressure Therapy, Postural Drainage.***)

Airways

Tubes that conduct outside air into the lungs. The lungs have many airways of varied sizes. The largest, or central, airway is the trachea. This branches into smaller airways called the bronchi. These divide into even smaller branches called bronchioles that end in alveoli. (Also see ***Alveoli, Bronchioles, Bronchus, Lower Respiratory Tract, Respiratory System, Trachea, Upper Respiratory Tract.***)

Alveoli
The millions of tiny air sacs in the lungs at the ends of bronchioles where oxygen is exchanged for carbon dioxide from the blood. In CF, mucus clogs alveoli and interferes with oxygen–carbon dioxide exchange. (Also see ***Airways, Lower Respiratory Tract, Mucus, Respiratory System.***)

Amniocentesis
A test used to find or "screen" for genetic defects in the fetus. A hollow needle is put through the mother's abdomen into the uterus. A small amount of amniotic fluid around the fetus is taken and tested. (Also see ***Amniotic Fluid, Chorionic Villus Sampling, Fetus, Gene***.)

Amniotic Fluid
The fluid around the fetus in the uterus. (Also see ***Amniocentesis, Fetus.***)

Antibiotic
A drug that can kill or slow the growth of bacteria, often used in CF to treat lung infections. (Also see ***Bacteria.***)

Antihistamine
A drug used to treat allergies that blocks histamine.

Anti-inflammatory
Reduces inflammation or swelling of body tissues. (Also see *Inflammation.*)

Anus
The lower opening of the digestive system where stool leaves the body. (Also see ***Digestive System, Stool.***)

Autogenic Drainage (AD)
An airway clearance technique that involves using various airflows to move mucus. (Also see ***Airway Clearance Techniques.***)

Autosomal-Recessive
A genetic trait or disorder that appears only when a person inherits a pair of chromosomes, each with the gene for the trait. One chromosome of the pair comes from the father and the other from the mother. Autosomal-recessive disorders occur only if each parent either is a carrier of the trait or has the trait. CF is autosomal-recessive. (Also see ***Base Pairs, Carrier, Chromosome, Gene, Genetic, Inherited, Hereditary.***)

B

Bacteria

Tiny one-celled organisms that are often the cause of infections. People with CF are prone to bacterial lung infections (often caused by *Staphylococcus aureus* or *Pseudomonas aeruginosa*). However, some bacteria normally found in the body are helpful. For example, *Escherichia coli* lives in the intestines and helps with digestion. (Also see ***Antibiotic, Pseudomonas aeruginosa, Intestine, Staphylococcus aureus.***)

Base Pairs

Small building blocks that make up genes. (Also see ***Autosomal-Recessive, Carrier, Chromosome, Gene, Genetic, Heredity, Inherited.***)

Basic Defect

In CF, the CFTR gene mutation makes faulty CFTR protein. (Also see ***Cystic Fibrosis Transmembrane Conductance Regulator.***)

Bile

A liver secretion that helps digestion. (Also see ***Digestion, Secretion.***)

Biotech

Short for ***biotechnology.*** Applying biological science to technology, including drug development. (Also see ***Research***.)

Blocked Intestine

The inability of food to move from the stomach through the intestine and out of the body.

Blood Pressure

Force exerted by the heart pumping blood.

Bronchiectasis

Chronic condition when the bronchi of the lungs are stretched or dilated beyond their normal dimensions. This leads to abnormal breathing and a productive cough. This can occur in CF. (Also see ***Bronchus, Chronic, Cough, Lower Respiratory Tract, Respiratory System.***)

Bronchioles

The smallest airways of the lungs leading from the bronchi to the alveoli. (Also see ***Airways, Alveoli, Bronchus, Lower Respiratory Tract, Respiratory System, Trachea.***)

Bronchitis
An inflammation of the bronchi caused by infection or exposure to cold or irritants. Symptoms include fever and cough. Many people with CF often have bronchitis. (Also see ***Bronchus, Symptom.***)

Bronchodilator
Medicine that opens bronchial tubes for easier breathing and to clear mucus, often given to relieve bronchospasm. (Also see ***Bronchioles, Bronchus, Mucus.***)

Bronchus/Bronchi
The large airways that move air from the trachea to the lungs. The bronchi branch into smaller airways called bronchioles. These lead to the alveoli. In CF, mucus clogs the bronchi and interferes with breathing. (Also see ***Airways, Alveoli, Bronchiectasis, Bronchioles, Lower Respiratory Tract, Mucus, Respiratory System, Trachea.***)

Buccal Smear
Cells brushed from the inside of the cheek. (Also see ***Cell.***)

Burkholderia Cepacia Complex
A group of bacteria called *B. cepacia* for short. In CF, *B. cepacia* can cause lung infection. *B. cepacia* may be spread from person to person with CF.

C

Carrier
A person having a single gene for a genetic trait or disorder such as CF. Carriers show no signs of the disease. In CF, each parent of a child with CF either has CF or is a CF carrier. (Also see ***Autosomal-Recessive, Gene, Genetic, Heredity, Inherited.***)

Cell
The basic unit of living organisms.

Cervical
Having to do with the cervix. (Also see ***Cervix, Reproductive System.***)

Cervix
The opening of the uterus. (Also see ***Uterus, Reproductive System.***)

CFRD
See ***Cystic Fibrosis–Related Diabetes***.

CFTR

See ***Cystic Fibrosis Transmembrane Conductance Regulator.***

Chest Physical Therapy (Chest PT or CPT)

An airway clearance technique that often includes both postural drainage and percussion. Helps to loosen mucus in the lungs. (Also see ***Airway Clearance Techniques, Percussion, Postural Drainage.***)

Chorionic Villus Sampling

A test to find gene defects in the fetus. A small piece of placenta is taken and tested early in pregnancy. A thin tube is put through the mother's vagina and cervix, or a thin needle is inserted through the abdomen into the uterus. (Also see ***Amniocentesis, Fetus, Placenta.***)

Chromosome

The threadlike material that carries genes, the units of heredity. Chromosomes are in the nucleus of every living cell. Every person should have 23 pairs of chromosomes in each cell. (Also see ***Cell, Gene, Genetic, Inherited, Heredity.***)

Chronic

A disease or condition that lasts and is persistent — not acute, which is characterized by a sudden onset, sharp rise and short course. CF is a chronic disease.

Cilia

Hairlike structures found on the surface of many cells in the body. In the respiratory system, cilia line the airways. They move together to push mucus to the trachea (windpipe) where it can be coughed up or huffed out or swallowed. Thick mucus, infection, cigarette smoke and other irritants can slow cilia and hinder this natural cleaning mechanism. (Also see ***Airways, Cough, Huffing, Mucus, Respiratory System, Tissue, Trachea.***)

Cirrhosis

Scarring of the liver; this can be caused by many diseases. Occurs in about 1 percent of people with CF. This is caused by thick secretions blocking the bile ducts. (Also see ***Duct, Secretion.***)

Clubbing

Rounded, enlarged tips of the fingers and toes. Clubbing often represents a chronic shortage of oxygen in the blood. Occurs in CF, congenital heart disease and other heart, lung and gastrointestinal diseases. (Also see ***Chronic, Sign.***)

Compressor
See ***Air Compressor.***

Conception
When a man's sperm joins with a woman's egg to form an embryo. (Also see ***Embryo, Fertilization, Fetus.***)

Contagious
Able to be spread from person to person, like an illness. CF is not contagious!

Contraception
Methods used to prevent pregnancy.

Cor Pulmonale
Enlargement of the right side of the heart. This happens when the heart has to work harder to get blood through the lungs. It can lead to right-sided heart failure. Cor pulmonale can occur in CF.

Cough/Coughing
A normal way for the body to clear the respiratory system of irritating and harmful things such as smoke, gases, dust and increased mucus. (Also see ***Respiratory System, Mucus.***)

Culture Medium
What sputum is put on for germs to grow in the laboratory, so the germ can be identified. (Also see ***Sputum***.)

Cystic Fibrosis–Related Diabetes (CFRD)
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. (Also see ***Diabetes, Glucose.***)

Cystic Fibrosis Transmembrane Conductance Regulator (CFTR)
A protein that makes the channel in the cell where chloride moves in and out.

D

Debris
Remains of something. For inhaled medicine, debris could be medication or sputum.

Dehydrate/Dehydration
Loss of too much water.

Diabetes (Diabetes Mellitus)

A pancreatic problem causing abnormal insulin production. Diabetes affects how the body uses sugar and other foods. It is treated by diet changes (less sugar intake) and insulin. (Also see *Pancreas, Cystic Fibrosis–Related Diabetes.*)

Diagnose

To find the cause of health problems.

Diaphragm

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

Diarrhea

Frequent, loose stools. (Also see *Stool.*)

Digestion/Digest

The process of breaking down the food we eat and absorbing its nutrients into the body for energy. (Also see *Absorption, Digestive System.*)

Digestive System

The organs that take in, digest and get rid of food. Includes the mouth, salivary glands, throat (pharynx), esophagus, stomach, intestines, liver, pancreas, colon, rectum and anus. In CF, thick mucus blocks some passages in the digestive system, including the passage between the pancreas and intestines. (Also see *Anus, Digestion, Duct, Esophagus, Intestine, Mucus, Pancreas, Rectum.*)

DIOS

See *Distal Intestinal Obstructive Syndrome.*

Direct Contact Transmission

Spreading germs by touching someone's body.

Disinfect

To kill most of the germs.

Distal Intestinal Obstructive Syndrome (DIOS)

Partially or completely blocked intestine by abnormal stool. Occurs in older infants, children and adults with CF. (Also see *Intestine, Meconium, Meconium Ileus Equivalent.*)

Diuretics

Medicines that help remove excess fluid from the body.

DNA
Deoxyribonucleic acid. The chemical coding for a gene. DNA decides the "genetic message" in each cell, organ and organism. (Also see ***Cell, Chromosome, Gene, Genetic, Inherited, Heredity.***)

Droplet
A tiny drop of liquid from the mouth, nose or lungs.

Dry Powder Inhaler (DPI)
A device that delivers a very fine powder of medicine that a person breathes in.

Duct
A tube or passageway for secretions. Ducts are found in organs, organ systems and exocrine glands. In CF, thick mucus can block these ducts. (Also see ***Exocrine Glands, Mucus, Pancreas, Secretion.***)

E

Egg
A cell from the woman containing half of the chromosomes needed to form an embryo. When it unites with a sperm from the man, which also contains half of the chromosomes, an embryo is created. (Also see ***Cell, Chromosome, Embryo, Reproductive System.***)

Embryo
An unborn child, from conception to 3 months after conception. (Also see ***Conception, Fertilize, Fetus, In Vitro Fertilization.***)

Endocrine Glands
Ductless glands that make hormones that pass into the blood. Includes pituitary, adrenal and thyroid glands. These are not affected in CF. (Also see ***Duct, Hormone.***)

Endocrinologist
A doctor who specializes in treating the endocrine system or endocrine glands. (Also see ***Endocrine Glands.***)

Enzymes
Proteins that help make and increase certain chemical processes in the body, such as the breaking down of foods in digestion. Because people with CF have mucus that often blocks the passageways through which digestive enzymes from the pancreas flow, they may need enzyme replacements to digest food. (Also see ***Absorption, Digestive System, Mucus, Pancreas.***)

Epididymis
Part of the male reproductive system — a tube that stores and carries sperm from the testes to the vas deferens. (Also see ***Microsurgical Epididymal Sperm Aspiration, Percutaneous Epididymal Sperm Aspiration, Reproductive System, Testes, Vas Deferens.)***

Esophagus
The tube that leads from the throat (pharynx) to the stomach. (Also see ***Digestive System***.)

Evidence-Based
The information is based on strong and reliable data from research.

Exacerbation
Signs and symptoms that show a need for treatment. (Also see ***Sign, Symptom.***)

Exhaling
Breathing out; the flow of air out of the lungs with each breath.

Exocrine Glands
Glands that normally make thin, slippery secretions including sweat, mucus, tears, saliva and enzymes. These secretions move through ducts (small tubes) to the surface of the body or into hollow organs, such as intestines or airways. CF affects these glands. Their ducts can be blocked by mucus. (Also see ***Airways, Duct, Intestine, Mucus, Secretion.***)

F

Fallopian Tubes
The tubes that carry the egg from the ovaries to the uterus. (Also see ***Egg, Ovaries, Reproductive System, Uterus.***)

False-Negative
Test result that incorrectly shows the absence of a disease or condition. A false-negative sweat test would show that the person does not have CF when they actually do have CF. (Also see ***Sweat Test.***)

False-Positive
Test result that incorrectly shows the presence of a disease or condition. A false-positive sweat test would show that the person has CF when they do not have CF. (Also see ***Sweat Test.)***

Fertile/Fertility

The ability to conceive and have children. (Also see ***Conception, Fertilize.***)

Fertilize/Fertilization

When a man's sperm unites with a woman's egg to form an embryo. (Also see ***Conception, Embryo, Fetus, In Vitro Fertilization.***)

Fetus

An unborn child, from 3 months after conception to birth. (Also see ***Conception, Embryo.***)

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 helps digest food in the intestines.

Gallstone

A stonelike mass in the gallbladder or bile duct. (Also see ***Duct.***)

Gastroenterologist

A doctor who specializes in treating the digestive system. (Also see ***Digestive System, Gastrointestinal.***)

Gastrointestinal (GI)

Pertaining to the stomach and intestine, or digestive system. (Also see ***Digestive System, Intestine.***)

Gene

The main unit of heredity. Each chromosome carries hundreds of genes. Genes decide body traits such as eye and hair color, height and facial features and also many health problems. CF is caused by a mutation of a gene. A child inherits CF at conception, when two CF genes are received, one from each parent. (Also see ***Autosomal-Recessive, Base Pairs, Carrier, Chromosome, Genetic, Hereditary, Inherited, Mutation.***)

Genetic

Hereditary or inherited. (Also see ***Autosomal-Recessive, Base Pairs, Carrier, Chromosome, Gene, Hereditary, Inherited.***)

Germs

Organisms that can cause infection. This includes bacteria and viruses. (Also see ***Bacteria, Virus.***)

GI

See ***Gastrointestinal.***

Glucose

A type of sugar that is found in many foods, and an important nutrient used by all cells for energy. Blood glucose levels are high if a person has diabetes. (Also see ***Cystic Fibrosis–Related Diabetes, Diabetes.***)

Gynecologist

A doctor who specializes in treating the female reproductive system. (Also see ***Reproductive System.***)

H

Hand Hygiene

A general term for cleaning hands; this includes the use of soap and water or an alcohol-based hand gel.

Heat Stroke

A health problem that occurs when the body gets too hot. The signs are weakness, nausea, dizziness and profuse sweating. It can occur when it is hot outside or in an extra-warm room.

Hemoptysis

Coughing up blood, often with sputum, because of broken small blood vessels in the lungs. Can occur in CF. (Also see ***Sputum.)***

Hereditary/Heredity

Traits or conditions that are genetically passed from parents to their children. (Also see ***Autosomal-Recessive, Base Pairs, Carrier, Chromosome, Gene, Genetic, Inherited.)***

Heterozygous

Someone who has inherited two different genes for a trait or disease. A person who is a heterozygote for CF has two different mutations in the genes for CF. (Also see ***Autosomal-Recessive, Base Pairs, Carrier, Chromosome, Gene, Genetic, Hereditary, Inherited, Mutation.)***

High-Frequency Chest Wall Oscillation

An airway clearance technique that involves wearing an inflatable vest attached to a machine that vibrates rapidly to loosen mucus in the lungs. (Also see ***Airway Clearance Technique, Mucus.***)

Homozygous

Someone who has inherited two of the same genes for a certain trait or disease. Someone with CF who is homozygous for CF has two of the same mutation of the CF gene. (Also see ***Autosomal-Recessive, Base Pairs, Carrier, Chromosome, Gene, Genetic, Hereditary, Inherited, Mutation.)***

Hormone

Secretion from the endocrine glands. Hormones regulate body functions including growth, maturation and heart rate. (Also see *Endocrine Glands, Secretion.*)

Huffing/Huff

An airway clearance technique using a gentle type of cough. Huffing is done by tightening the stomach muscles while forcefully pushing air out with the mouth open. It is like what we do when we "huff" onto a mirror or window to fog it up. (Also see ***Airway Clearance Techniques, Cough.)***

I

ICSI

See *Intracytoplasmic Sperm Injection.*

IDEA

See *Individuals with Disabilities Education Act.*

IEP

See *Individualized Education Plan.*

Immunization

Vaccination or "shot" to help the body build a defense against an illness. (Also see ***Vaccination.***)

Immunoreactive Trypsinogen (IRT) Test

A blood test used 2 or 3 days after birth that may be done with DNA tests to find out if a baby has CF. Immunoreactive trypsinogen, or IRT, is a chemical normally found in small amounts in the body. People with CF tend to have high levels of IRT.

Indirect Contact Transmission

Spreading germs by touching something that another person has touched (such as a doorknob or cup) with any part of the body.

Individualized Education Plan (IEP)
A plan between a child's family and a school to help meet the child's learning needs. The plan tells how the school will manage things such as absences or medical treatments at school, such as taking enzymes.

Individuals with Disabilities Education Act (IDEA)
A federal law that makes public elementary and high schools give free and appropriate education to children with disabilities.

Infertile/Infertility
Unable to get pregnant (female), or to cause pregnancy (male).

Inflammation/Inflammatory
The swelling of body tissues because of irritation or injury. Inflammation occurs with an infection.

Inhalation
Breathing in; the flow of air into the lungs.

Inherit/Inherited
Traits or conditions that are genetically passed from parents to their children. (Also see ***Autosomal-Recessive, Base Pairs, Carrier, Chromosome, Gene, Genetic, Heredity.***)

Insulin
A hormone, made by the pancreas, that helps sugar move from the blood into the cells. Sugar is fuel for the body to grow and fight infection.

Intestinal
Having to do with the intestine. (Also see ***Digestive System, Gastrointestinal, Intestine.)***

Intestine
Tube in the digestive system that connects the stomach to the anus. The long, narrow, upper part is the small intestine. The short, wide, lower part is the large intestine. (Also see ***Anus, Digestive System.***)

Intracytoplasmic Sperm Injection (ICIS)
Fertilization of an egg by injecting a sperm into it during the process of in vitro fertilization. (Also see ***Egg, Fertilization, In Vitro Fertilization, Sperm.***)

Intrapulmonary Percussive Ventilation (IPV)
An airway clearance technique that involves breathing through a device that creates a rapid vibration to loosen the mucus in the lungs. (***Also see Airway Clearance Technique, Mucus.***)

Intravenous (IV)
Putting a medicine right into a blood vessel, usually a vein, using a thin needle and a tube.

In Vitro Fertilization
Mixing sperm with an egg outside the woman's body. The fertilized egg is then put into the woman's uterus. (Also see *Conception, Egg, Embryo, Fertilization, Sperm, Uterus.*)

Ion Transport
The movement of sodium and chloride in and out of the cell is one type of ion transport.

IRT
See *Immunoreactive Trypsinogen Test.*

IV
See *Intravenous.*

L

Legislative
Having to do with making laws.

Lower Respiratory Tract
The airways and the lungs (trachea, bronchi, bronchioles and alveoli). (Also see *Alveoli, Bronchioles, Bronchus, Respiratory System, Trachea.*)

M

Malabsorption
Poor uptake of nutrients from food for use by the body. In CF, mucus may plug ducts of digestive organs and block the secretion of enzymes and hormones. This makes many nutrients unavailable for use in body maintenance and growth. (Also see *Absorption, Digestion, Digestive System, Duct, Enzyme, Hormone, Mucus, Pancreas, Secretion.*)

MDI
See *Metered Dose Inhaler.*

Meconium
The first newborn stool, often passed within a few hours after birth. It contains mucus and other secretions. (Also see *Meconium Ileus, Mucus, Secretion.*)

Meconium Ileus

Blockage of the intestines of a newborn with very thick meconium. It can be the earliest symptom of CF, and occurs in 7 percent to 10 percent of people with CF. (Also see ***Intestine, Meconium, Symptom.***)

Meconium Ileus Equivalent (Distal Intestinal Obstructive Syndrome [DIOS])

Partially or completely blocked intestine by abnormal stool. It occurs in older infants, children and adults with CF. (Also see ***Distal Intestinal Obstructive Syndrome, Intestine, Meconium, Meconium Ileus.***)

MESA

See ***Microepididymal Sperm Aspiration.***

Metered Dose Inhaler (MDI)

A device that helps a person to measure and inhale a certain amount of medicine.

Microepididymal Sperm Aspiration (MESA)

An outpatient surgical technique in which sperm are taken directly from a man's epididymis. For men with CF who are infertile, this method may help them conceive a biological child when combined with in vitro fertilization (and usually intracytoplasmic sperm injection). (Also see ***Epididymis, Intracytoplasmic Sperm Injection, In Vitro Fertilization, Percutaneous Epididymal Sperm Aspiration, Testicular Sperm Extraction***.)

Mucolytics

Medicines that thin mucus, making it easier to cough out. (Also see ***Mucus.***)

Mucous Membrane

Tissue that contains mucus-making glands. Mucous membranes are found in the nose, mouth, lungs, esophagus, stomach and intestines. (Also see ***Esophagus, Intestine, Mucus, Tissue.***)

Mucous Plugs

Thick mucus in a duct or airway that can block the flow of secretions or air. (Also see ***Airway, Duct, Mucus, Pancreas, Secretion.***)

Mucus

A fluid made by mucous membranes and glands. Mucus is normally thin and slippery. In CF, the mucus is often thick and sticky. (Also see ***Mucous Membrane, Phlegm, Sputum.***)

Mutation(s)

A change in a gene that lasts forever. (Also see ***Autosomal-Recessive, Base Pairs, Carrier, Chromosome, Gene, Genetic, Hereditary, Inherited.***)

N

Nasal Polyps

Small growths of swollen mucous membrane that project into the nasal passages. They are common in people with CF, and are often multiple or recurrent. Nasal polyps can be surgically removed. (Also see ***Mucous Membrane.***)

Nebulizer

A device that makes and delivers a medicine mist when attached to an air compressor. (Also see ***Aerosol, Air Compressor, Antibiotics, Mucolytics.***)

Nervous System

The part of the body that includes the brain, spinal cord and nerves.

Nucleus

The center or "brain" of the cell, containing the chromosomes. (Also see ***Cell, Chromosome.***)

O

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. (Also see ***Cystic Fibrosis–Related Diabetes, Diabetes, Glucose***)

Oscillating Positive Expiratory Pressure (Oscillating PEP)

An airway clearance technique that involves breathing out through a device many times in a row to loosen mucus in the lungs. (Also see ***Airway Clearance Technique, Mucus.***)

Osteopenia

When bones have fewer minerals and are weak.

Osteoporosis

When bones are less thick or dense and are weak. People with osteoporosis have a higher risk for bone breaks (fractures).

Ovaries

The female organs where hormones and eggs are made. (Also see *Hormone, Egg, Reproductive System.*)

Ovulate

To release an egg into the fallopian tube. (Also see *Egg, Fallopian Tube, Ovaries, Reproductive System.*)

P

Pancreas

A long organ with glands found behind the stomach. The pancreas secretes enzymes through ducts into the intestine to break down food. In CF, mucus may obstruct the pancreatic ducts, preventing digestion. Another part of the pancreas has endocrine tissue, which makes the hormone insulin. Insulin controls storage and oxidation of sugar. (Also see *Cystic Fibrosis–Related Diabetes, Digestive System, Digestion, Duct, Endocrine Glands, Enzyme, Exocrine Glands, Hormone, Intestine, Mucus, Tissue.*)

Pancreatic

Having to do with the pancreas. (Also see *Pancreas.*)

Penis

The male organ through which urine and semen leave the body. (Also see *Reproductive System, Semen.*)

PEP

See *Positive Expiratory Pressure.*

Percussion

An airway clearance technique that involves clapping, with a cupped hand, and vibrating the chest to loosen mucus in the lungs. (Also see *Airway Clearance Technique, Chest Physical Therapy, Mucus.*)

Percutaneous Epididymal Sperm Aspiration (PESA)

An outpatient surgical technique in which sperm are taken directly from a man's epididymis. For men with CF who are infertile, this method may help them conceive a biological child when combined with in vitro fertilization (and usually intracytoplasmic sperm injection). (Also see *Epididymis, Intracytoplasmic Sperm Injection, In Vitro Fertilization, Microepididymal Sperm Aspiration, Testicular Sperm Extraction.*)

PESA

See *Percutaneous Epididymal Sperm Aspiration.*

PFTs

See *Pulmonary Function Tests.*

Phlegm

Mucus made from glands in the lungs and airways. (Also see *Airways, Mucus, Secretion, Sputum.*)

Pilocarpine Iontophoresis

A test that causes sweating by putting the chemical pilocarpine on a small area of the skin, which then receives a small electrical current. Quantitative pilocarpine iontophoresis is the usual way to do a sweat test to diagnose CF. (Also see *Sweat Test.*)

Placenta

What an unborn child gets oxygen and nutrition from through the mother.

Pneumonia

An inflammation of the lungs often caused by a bacterial or viral infection. Pneumonia is a problem in people with CF. (Also see *Bacteria, Virus.*)

Pneumothorax

A sudden, partial or complete lung collapse caused by a break in lung tissue or an airway, which lets air escape from the lung and get trapped between the lung and chest wall. It can occur in people with CF.

Positive Expiratory Pressure (PEP) Therapy

An airway clearance technique that involves breathing through a mouthpiece or mask connected to a prescribed resistor. (Also see *Airway Clearance Technique.*)

Postural Drainage (PD or Bronchial Drainage)

An airway clearance technique that involves lying in various positions to drain mucus from the lungs. (Also see *Airway Clearance Technique, Chest Physical Therapy, Mucus, Percussion.*)

Prenatal/Prenatally

Refers to the time between conception and birth. (Also see *Conception.*)

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.

Prostate Gland
A gland in the male body that makes semen. (Also see *Reproductive System, Semen.*)

Pseudomonas aeruginosa
A type of bacteria that often lives in the lungs of people with CF and causes lung infections. (Also see *Antibiotic, Bacteria.*)

Pulmonary
Relating to the lungs.

Pulmonary Function Tests (PFTs)
Tests to check lung function. Along with patient history and examination, PFTs help doctors diagnose, plan therapy and determine response to therapy. They can be used with children aged 5 years and older. PFTs measure air flow and lung volumes.

Pulmonologist
A doctor who specializes in treating the respiratory system. (Also see *Respiratory System.*)

R

Rectal Prolapse
When the inner lining of the rectum comes out (prolapses) through the anus. This may occur in children with CF because of digestion problems. CF is the most common cause of rectal prolapse in infants and children in the United States. (Also see *Anus, Digestion, Rectum.*)

Rectum
The last part of the large intestine joining the colon to the anus. (Also see *Anus, Digestive System, Intestine.*)

Reproductive System
In the male, includes the testes, sperm, vas deferens, prostate gland, semen, urethra, scrotum and penis. In the female, includes the eggs, ovaries, fallopian tubes, uterus, cervix and vagina. (Also see *Cervix, Egg, Fallopian Tubes, Ovaries, Penis, Prostate Gland, Semen, Sperm, Testes, Uterus, Vagina, Vas Deferens.*)

Research
- **Applied Research:** Studies that apply basic research findings to problems including diseases and symptoms. Creating new respiratory equipment or studying cell defects in the sweat glands of people with CF are applied research. (Also see *Cell, Symptom.*)

- **Basic Science Research:** Studies that increase knowledge of basic life processes. To learn more about CF, scientists do basic science studies such as gene studies and research on how cells work. (Also see ***Cell, Gene, Genetic.***)
- **Clinical Research:** Studies that improve diagnosis and treatment. Studies on medicines, lung function testing, nutrition and sweat testing methods are examples. (Also see ***Diagnose, Pulmonary Function Tests, Sweat Test.***)

Respiratory Syncytial Virus (RSV)

A virus that can cause severe respiratory infections, especially in young children and the elderly.

Respiratory System

The part of the body that includes all structures that air moves through while breathing. Also includes the pleura, ribs and intercostal muscles that support these structures. The upper respiratory tract includes the nose, sinuses, throat (pharynx and larynx). The lower respiratory tract includes the trachea, bronchus, bronchioles and alveoli. In CF, thick mucus clogs parts of the respiratory system. (Also see ***Trachea, Bronchus, Bronchioles, Alveoli, Sinuses.***)

S

Saline

A mixture of salt and water similar to the body's normal tissue fluids. (Also see ***Tissue.***)

Secretion

A product of a gland, such as sweat or saliva. (Also see ***Endocrine Gland, Exocrine Gland.***)

Section 504 of the Rehabilitation Act of 1973

A federal law that prohibits discrimination against a person because of a disability by any group that gets federal funds. This law applies to day care and the public and private schools that get federal funds.

Semen

A sticky, white fluid of the male reproductive system that contains the sperm. (Also see ***Prostate Gland, Reproductive System, Sperm.***)

Sexually Transmitted Diseases (STDs)

Diseases that can be caught through sexual contact.

Sibling(s)

Brothers and sisters.

Sign
The clues of an illness or problem that you can see or measure, such as fingertip clubbing, nasal polyps or rectal prolapse. (Also see ***Clubbing, Nasal Polyps, Rectal Prolapse, Symptom.***)

Sinuses
The air spaces in the bones of the skull, mostly connected with the nose. (Also see ***Respiratory System, Sinusitis.***)

Sinusitis
An inflammation of the lining of the sinuses that causes fluid to drain into the nasal cavity. (Also see ***Sinuses, Respiratory System.***)

Sperm
A cell from the male containing half of the chromosomes needed to form an embryo. When it unites with an egg from the woman, which also contains half of the chromosomes, an embryo is created. (Also see ***Cell, Chromosome, Embryo, Reproductive System.***)

Sputum
Mucus or phlegm coughed up from lungs. (Also see ***Mucus, Phlegm.***)

Sputum Culture
A test to see what germs are growing in the sputum.

Staphylococcus aureus (Staph)
A type of bacteria that can cause infections. In CF, staph often causes lung infections. It is treated with antibiotics. (Also see ***Antibiotic, Bacteria.***)

Sterile/Sterility
The absence of sperm, the male sex cell. Men with CF are usually not sterile, but are very often infertile. (Also see ***Infertile/Infertility, Sperm.***)

Stool
Another word for bowel movement, poop or feces.

Sweat Test
The test to diagnose CF. Measures the salt (sodium and chloride) in sweat. (Also see ***Diagnose, Pilocarpine Iontophoresis.***)

Symptom
A sign of a person's disease or condition. Something felt by the person. CF symptoms include persistent cough; wheeze; shortness of breath; bulky, very foul-smelling stools; and stomachache. (Also see ***Cough, Sign, Wheeze.***)

T

TESE
See ***Testicular Sperm Extraction***.

Testes
The round organs in the scrotum where hormones and sperm are made. Sometimes called testicles. (Also see ***Hormone, Reproductive System, Sperm.***)

Testicular Sperm Extraction (TESE)
An outpatient surgical technique in which sperm are taken directly from a man's testes. For men with CF who are infertile, this method may help them conceive a biological child when combined with in vitro fertilization (and usually intracytoplasmic sperm injection). (Also see ***Intracytoplasmic Sperm Injection, In Vitro Fertilization, Microepididymal Sperm Aspiration, Percutaneous Epididymal Sperm Aspiration, Testes***.)

Tissue
A group of cells of a similar type and function. (Also see ***Cell.***)

Trachea
The windpipe, or largest central airway. Connects the upper respiratory tract to the lower respiratory tract. (Also see ***Airways, Lower Respiratory Tract, Respiratory System, Upper Respiratory Tract.***)

U

Upper Respiratory Tract
The nose, sinuses, throat (pharynx and larynx). (Also see ***Respiratory System, Sinuses.***)

Uterus
A muscle sac (organ), also called the womb, where the baby grows until birth. (Also see ***Reproductive System.***)

V

Vaccination
Immunization or "shot" to help the body build a defense against an illness. (Also see ***Immunization.***)

Vagina
In women, the canal that leads from the uterus to the outside of the body. (Also see ***Reproductive System, Uterus.***)

Vas Deferens

A duct in the male reproductive system, which carries sperm from the testes to the prostate gland. In males with CF, this duct is often blocked, leading to sterility. (Also see ***Duct, Prostate Gland, Reproductive System, Sperm, Testes.***)

Virus

An organism, smaller than bacteria, which causes infections such as influenza, viral pneumonia, colds and hepatitis. (Also see ***Bacteria, Pneumonia.***)

Vitamins

Substances in foods needed for the body to function normally. When digestive enzymes are blocked in CF, vitamins from foods may be poorly absorbed by the body, so vitamin supplements are needed. (Also see ***Absorption, Digestive System, Malabsorption, Pancreas.***)

W

Wheeze/Wheezing

To breathe hard and with a whistling sound.

Appendix B: Resource List

In addition to the resources listed here, your cystic fibrosis (CF) care center has many resources available to share with you. Also, the Cystic Fibrosis Foundation's website, **www.cff.org**, has a wealth of fact sheets, answers to frequently asked questions, research updates and much more on a wide variety of CF-related topics and the latest breaking CF-related news.

If you choose to surf the Web for information related to CF, keep in mind that anyone can put information on the Internet. Although there is some good information online, there is also a great deal of information that is **not** correct. For tips on using the Internet to research health information, read the "Guide to 'Healthy' Web Surfing: Ways to Evaluate the Quality of Health Information on Web Sites" available from your CF care center or at **www.cff.org**.

Cystic Fibrosis Foundation:

www.cff.org
info@cff.org
1-800-FIGHT CF or 301-951-4422
6931 Arlington Road
Bethesda, MD 20814

The CF Foundation's website has information for adults and children with CF as well as parents and families related to CF care, CF care centers, the Patient Registry Report, advocacy, insurance and more.

Book:

Orenstein DM. *Cystic Fibrosis: A Guide for Patient and Family*, 4th ed., New York, NY: Lippincott Williams & Wilkins; 2012.

Support Groups:

Contact your local CF care center. (See Appendix E)

Cystic Fibrosis Family Education Program Publications:
The following resources are available from your local CF care center.

- *Beginning CF Care*
- *Managing Nutrition & Digestive Problems*
- *Managing Lung & Other Respiratory Problems*
- *Becoming a CF Manager*
- *Working With Your Child*
- *Lisa and Jason: Family, Friends, and Everyday Life: For Young Children With Cystic Fibrosis and Their Families* (stories and activities for young children)

CF Roundtable:
www.cfroundtable.com
248-349-4553
cfroundtable@usacfa.org
USACFA
PO Box 1618
Gresham, OR 97030

Newsletter for adults with CF from the United States Adult Cystic Fibrosis Association Inc. (USACFA).

MedlinePlus:
www.medlineplus.gov

MedlinePlus is the National Institutes of Health's website for patients and their families and friends. Produced by the National Library of Medicine, it provides information about diseases, conditions and wellness issues. MedlinePlus offers free, reliable, up-to-date health information.

National Institutes of Health*:*
www.clinicaltrials.gov

Website to find clinical trials.

Centers for Disease Control and Prevention (CDC):
www.cdc.gov

The CDC works to create the information and tools that people and communities need to protect their health — through health promotion; prevention of disease, injury and disability; and more.

American Lung Association:
www.lungusa.org
1-800-548-8252

Medicare:

www.medicare.gov
1-800-MEDICARE

Medicare provides health care coverage for people with long-term disabilities (and most Americans ages 65 years and older).

Social Security Disability Insurance (SSDI):

www.ssa.gov/disability
1-800-772-1213

The SSDI program provides income to people who are no longer able to work because of a disability; benefits are based on a person's work history. People who receive SSDI are eligible for Medicare after 2 years.

State Children's Health Insurance Program (SCHIP):

www.insurekidsnow.gov
1-877-KIDS-NOW (1-877-543-7669)

SCHIP provides health care coverage to uninsured children in families with incomes that are too high to qualify for Medicaid.

Supplemental Security Income (SSI):

www.ssa.gov/pubs/11000.html
1-800-772-1213

The SSI program provides a monthly stipend to people with disabilities (and low-income Americans ages 65 years and older) to help meet basic needs for food, shelter and clothing. People who receive SSI are often eligible for Medicaid.

CF Services Pharmacy Inc.:

www.cfservicespharmacy.com
1-800-541-4959

Cystic Fibrosis Services Inc. is a Walgreens alliance partner and is affiliated with the CF Foundation. A national mail-order pharmacy, CF Services is the nation's leading provider of specialty CF drugs, case management services and reimbursement support for people with CF.

CF Patient Assistance Foundation:

www.cfpaf.org
1-888-315-4154

A nonprofit subsidiary of the CF Foundation, offers financial assistance for CF medications and devices by providing co-payment and co-insurance assistance for eligible medications and devices, insurance benefit verification, reimbursement support, case management, patient advocacy and referrals to alternative assistance programs.

CF Legal Information Hotline:

www.cff.org
CFLegal@cff.org
1-800-622-0385

The Hotline provides free information about the laws that protect the rights of people with CF. It serves as a resource for CF care centers, people with CF and their families.

Health care and insurance information:

www.healthcare.gov

From the U.S. Department of Health and Human Services.

State Insurance Commissioners:

www.naic.org
1-816-783-8500

For information on health insurance or filing a complaint, visit the National Association of Insurance Commissioners.

Organ Donation and Transplantation:

www.organdonor.gov

U.S. Department of Health and Human Services.

College Scholarships:

- The Cystic Fibrosis Scholarship Foundation:
  www.cfscholarship.org
- CFCareForward Scholarship:
  www.CFCareForwardScholarship.com
- The Elizabeth Nash Foundation Scholarship Program:
  www.elizabethnashfoundation.org
- Federal Student Aid Information Center, U.S. Department of Education, "The Student Guide":
  www.studentaid.ed.gov
  1-800-4-FED-AID (1-800-433-3243)
  PO Box 84
  Washington, DC 20044-0084

Appendix C: An Introduction to Postural Drainage and Percussion

Postural drainage and percussion (PD & P), also known as chest physical therapy (CPT), is a way to help people with cystic fibrosis (CF) breathe with less difficulty and stay healthy. PD & P uses gravity and percussion to loosen the thick, sticky mucus in the lungs so it can be removed by coughing. Unclogging the airways is key to keeping lungs healthy.

PD & P is easy to do using the techniques described here. For the person with CF, PD & P can be done by physical therapists (PTs), respiratory therapists (RTs), nurses, parents, family and even friends.

People with CF sometimes use other types of treatments, such as inhaled bronchodilators and antibiotics, to keep their lungs healthy. If ordered, bronchodilators should be taken **before** PD & P to open the airways, and inhaled antibiotics should be taken **after** PD & P so that the medicine gets to the infection better. Your care center doctor or therapist will help you figure out a routine that will work best for you or your child.

Know Your Lungs

Learning more about the respiratory system and its relationship to other organs in the body can help you to understand why PD & P treatments are effective.

Getting Rid of Mucus

The goal of PD & P is to clear mucus from each of the five lobes of the lungs by moving mucus into the larger airways so that it can be coughed out. The right lung is composed of three lobes: the upper lobe, the middle lobe and the lower lobe. The left lung is made up of only two lobes: the upper lobe and the lower lobe.

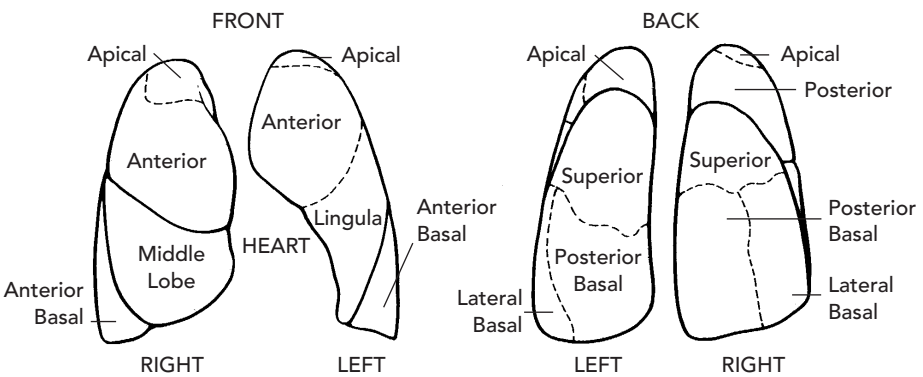


Figure 1: Anatomy of the Lungs

The lobes are divided into smaller sections called segments. The upper lobes on the left and right sides are each made up of three segments: top (apical), back (posterior) and front (anterior).

The lungs are made up of a network of air tubes, air sacs and blood vessels. These sacs allow for the exchange of oxygen and carbon dioxide between the blood and air. It is these segments that are being drained. Note the position of each lung segment in Figure 1 on the previous page.

Performing PD & P

The performance of PD & P involves a combination of techniques including multiple positions to drain the lungs, percussion, vibration, deep breathing and coughing.

Once the person is in one of the positions, the caregiver does percussion on the chest wall. This is usually given for a period of 3 to 5 minutes and sometimes followed by vibration over the same area for approximately 15 seconds (or during 5 exhalations). The person is then encouraged to cough or huff forcefully to get the mucus out of the lungs.

Description of PD & P Techniques

Postural drainage uses gravity to help move mucus from the lungs up to the throat. The person lies or sits in various positions so that the part of the lung to be drained is as high as possible. The part of the lung is then drained using percussion, vibration and gravity. For a complete description of these positions, see the diagrams later in this appendix. Your CF care team may tailor these positions to your or your child's needs.

Percussion or clapping by the caregiver on the chest wall over the part of the lung to be drained helps move the mucus into the larger airways. The hand is cupped as if to hold water but with the palm facing down as in Figure 2. The cupped hand curves to the chest wall and traps a cushion of air to soften the clapping.

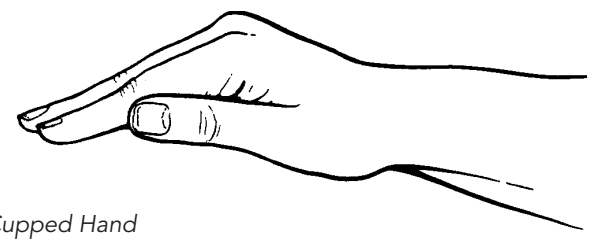


Figure 2: Cupped Hand

Percussion is done forcefully and with a steady beat. It should not be painful or sting if the hand is cupped properly. Each percussion also should have a hollow sound. Most of the movement is in the wrist with the arm relaxed, making percussion less tiring to do.

Special attention must be taken to not clap over the spine, breastbone, stomach and lower ribs or back to prevent injury to the spleen on the left, the liver on the right and the kidneys in the lower back.

Different devices may be used instead of the traditional cupped palm method for percussion. Ask your doctor or physical or respiratory therapist for advice.

Vibration gently shakes the mucus into the larger airways. The caregiver places a hand firmly on the chest wall over the part of the lung being drained and tenses the muscles of the arm and shoulder to create a fine shaking motion. Then, the caregiver applies a light pressure over the area being vibrated. (The caregiver also may place one hand over the other, then press the top and bottom hand into each other to vibrate.) Vibration is done with the flattened hand, not the cupped hand, as in Figure 3. Exhalation should be as slow and as complete as possible.

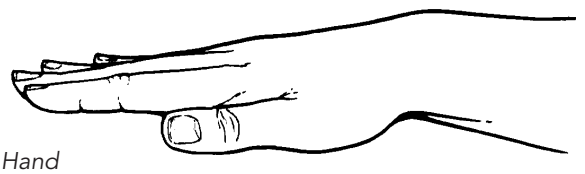


Figure 3: Flat Hand

Deep breathing moves the loosened mucus and may lead to coughing. Breathing with the diaphragm, belly breathing or lower chest breathing is used to help the person take deeper breaths and get the air into the lower lungs. The belly moves outward when the person breathes in and sinks in when he or she breathes out.

Coughing is key to clear the airways of mucus. A forced but not strained exhalation, following a deep inhalation, may help a person cough. The mucus can then be coughed out.

Huffing

Huffing is a type of cough. It also involves taking a breath in and actively exhaling. It is more like "huffing" onto a mirror or window to steam it up. It is not as forceful as a cough but may work better and be less tiring.

Timing of PD & P

Generally, each treatment session can last for 20 to 40 minutes. PD & P is best done before meals or 1½ to 2 hours after eating to decrease the chance of vomiting. Early morning and bedtimes usually are recommended. The length of PD & P and the number of times of day it is done may need to be increased if the person is more congested or getting sick. Your CF doctor or therapist will help you know what positions, how often and how long PD & P should be done.

Enhancing PD & P for the Person With CF and the Caregiver

Both the person with CF and the caregiver should be comfortable during PD & P. Before starting, the person with CF should remove tight clothing, jewelry, buttons and zippers around the neck, chest and waist. Light, soft clothing, such as a T-shirt, may be worn. **Do not do PD & P on bare skin.** The caregiver should remove rings and other bulky jewelry such as watches or bracelets. A supply of tissues or a place to cough out the mucus should be nearby.

Doing PD & P Comfortably and Carefully

The caregiver should not lean forward when doing percussion, but should remain in an upright position to protect his or her back. The table on which the person with CF lies should be at a comfortable height for the caregiver.

Pillows, sofa cushions, bundles of newspapers under pillows for support, cribs with adjustable mattress heights/tilts, foam wedges and bean bag chairs work for many. Infants can be positioned with or without pillows in the caregiver's lap.

Purchasing Equipment

Equipment such as drainage tables, electrical and nonelectrical palm percussors and vibrators may be helpful. These can be purchased from medical equipment stores. Older children and adults may find percussors useful when doing their own PD & P. Talk to your doctor or therapist at your CF care center about equipment for PD & P.

Making PD & P More Enjoyable

To enhance the quality of the time you spend doing PD & P, do one of the following:

- Schedule PD & P around a favorite TV show.
- Play favorite music or recorded stories.
- Spend time talking or singing before, during and after PD & P.
- For kids, encourage blowing or coughing games during PD & P, such as blowing pinwheels or coughing the deepest cough.
- Ask willing and capable relatives, friends, brothers and sisters to do PD & P. This can provide a welcome break from the daily routine.
- Minimize interruptions.

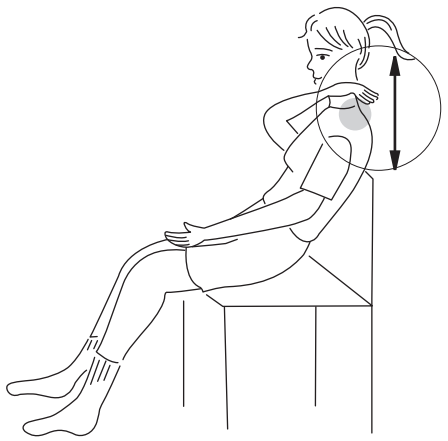
Finding ways that make PD & P more enjoyable can help you keep a regular routine and get maximum health benefits.

Instructions for Postural Drainage Positions

The following diagrams show and describe the positions for PD & P. The shaded areas show where the chest should be percussed or clapped.

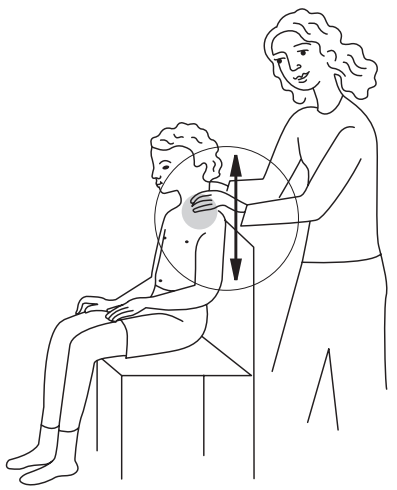
Pillows may be used for added comfort. If the person tires easily, the order of the positions can be varied, but all areas of the chest should be percussed or clapped.

Please remember to percuss and vibrate only over the ribs. Avoid percussing and vibrating over the spine, breastbone, stomach and lower ribs or back to prevent injury. Do not percuss or vibrate on bare skin.



Self-Percussion–Upper Lobes

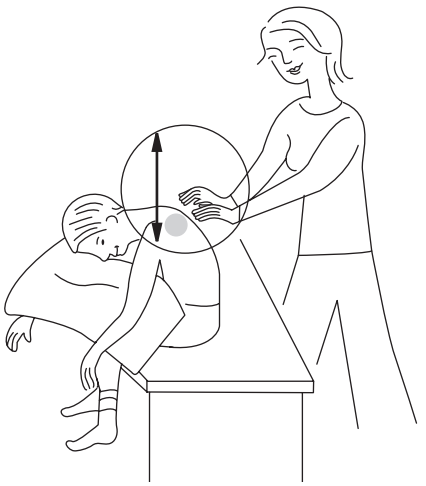
The person with CF should sit upright and reach across his or her chest to clap on front of chest over the muscular area between the collarbone and the top of the shoulder blade. Repeat on the opposite side.



Upper Front Chest–Upper Lobes

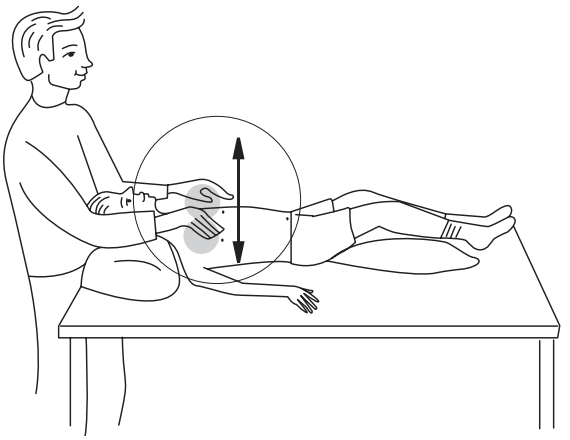
Have the person with CF sit upright. Clap on both sides of upper front chest over the muscular area between the collarbone and the top of the shoulder blade.

** The people with CF are shown without shirts to better demonstrate the PD & P technique in illustrations. Images are from the CF Family Education Program and used with permission from Baylor College of Medicine.*



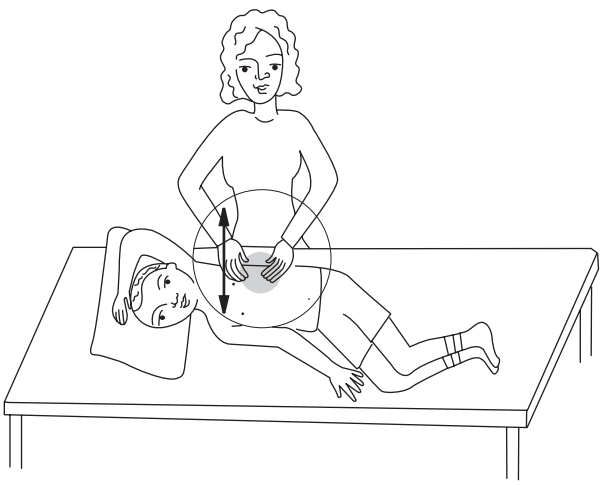
Upper Back Chest–Upper Lobes

Have the person with CF sit up and lean forward on a pillow over the back of a sofa or soft chair at a 30 degree angle. Stand or sit behind him or her and clap both sides of the upper back. Take care not to clap on the backbone.



Upper Front Chest–Upper Lobes

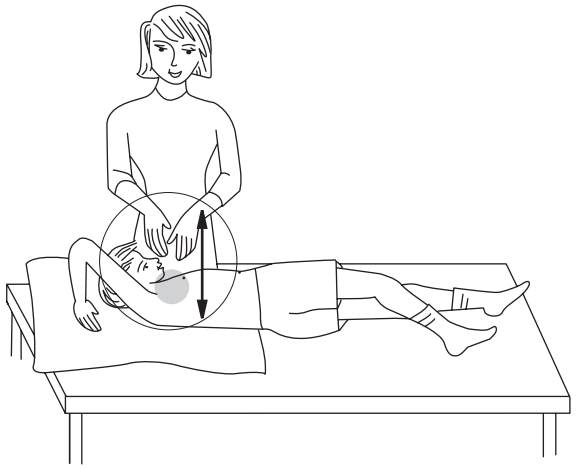
Have the person with CF lie on his or her back with arms to sides. Stand behind his or her head. Clap both sides of the chest between the collarbone and nipple.



Left Side Front Chest

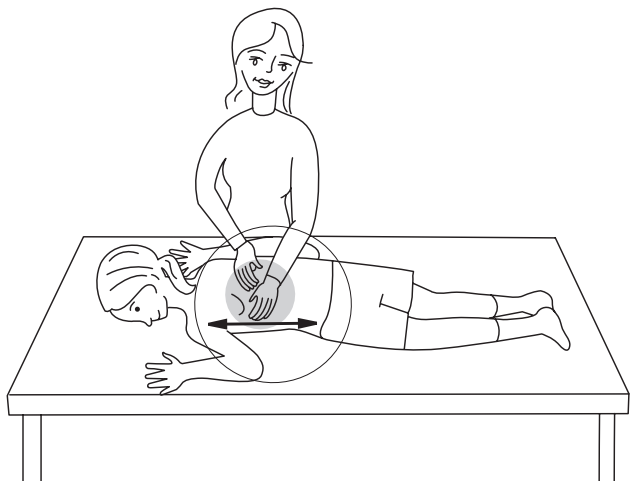
Have the person with CF lie with left side up and raise his or her left arm over head. Clap over the ribs just below the nipple area on front side of left chest. Do not clap on the stomach.

** The people with CF are shown without shirts to better demonstrate the PD & P technique in illustrations. Images are from the CF Family Education Program and used with permission from Baylor College of Medicine.*



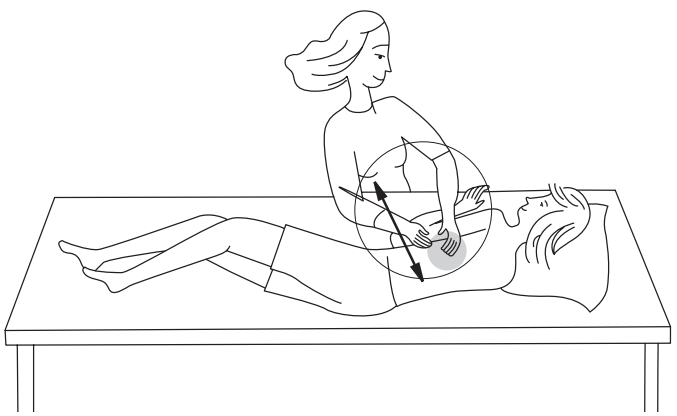
Right Side Front Chest

Have the person with CF lie with right side up and raise right arm over head. Clap over the chest just below the nipple area on front side of right chest. Do not clap lower ribcage.



Lower Back Chest–Lower Lobes

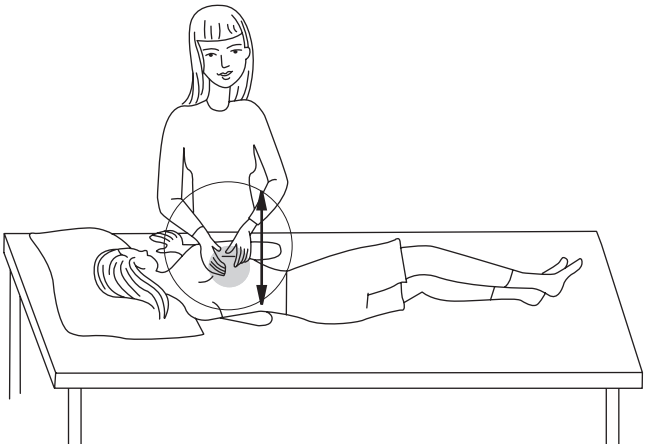
Have the person with CF lie on his or her stomach. Clap both sides at the bottom of chest just above the bottom edge of the ribcage. Do not clap lower ribcage or over the backbone.



Left Lower Side Back Chest–Lower Lobe

Have the person with CF lie with left side up and roll toward the caregiver a quarter turn so he or she can reach the back of the person with CF. Clap on lower left side of chest just above the bottom edge of the ribcage.

** The people with CF are shown without shirts to better demonstrate the PD & P technique in illustrations. Images are from the CF Family Education Program and used with permission from Baylor College of Medicine.*



Right Lower Side Back–Lower Lobe

Have the person with CF lie right side up and roll toward the caregiver a quarter turn so he or she can reach the back of the person with CF. Clap on lower right side of the chest just above the bottom edge of the ribcage.

** The people with CF are shown without shirts to better demonstrate the PD & P technique in illustrations. Images are from the CF Family Education Program and used with permission from Baylor College of Medicine.*

Appendix D: Daily Record of Diet, Treatments and Symptoms

Many people with CF and parents of children with CF find it helpful to keep daily records. The records on the following pages can help you keep track of any changes in health. You can copy these pages if you wish to keep such records, and bring them to clinic visits.







Food Diary

Date:_____

BREAKFAST: Start time_____ End time_____

Food and drinks (type/amount):

Number of enzymes:

Time taken:

Comments:

MORNING SNACK: Start time_____ End time_____

Food and drinks (type/amount):

Number of enzymes:

Time taken:

Comments:

LUNCH: Start time_____ End time_____

Food and drinks (type/amount):

Number of enzymes:

Time taken:

Comments:

AFTERNOON SNACK: Start time_____ End time_____

Food and drinks (type/amount):

Number of enzymes:

Time taken:

Comments:

DINNER: Start time_____ End time_____

Food and drinks (type/amount):

Number of enzymes:

Time taken:

Comments:

EVENING SNACK: Start time_____ End time_____

Food and drinks (type/amount):

Number of enzymes:

Time taken:

Comments:

Treatment Diary

Date:_____

AIRWAY CLEARANCE

Type(s):	Time(s):	How long?	Comments:

INHALED MEDICATIONS

Name(s) of medication:	Time(s) taken:	Dosage(s):	Comments:

ENZYMES

Name of enzyme:	Times taken:	Dosages:	Comments:

VITAMINS

Name of vitamin:	Time(s) taken:	Dosage(s):	Comments:

OTHER MEDICATIONS

Name of medication(s):	Time(s) taken:	Dosage(s):	Comments:

Symptom Diary

Date:_____

COUGH AND BREATHING

When does coughing occur?	How does the cough sound? (e.g., Does it sound wet?)	Amount of sputum coughed up, color and thickness	Any wheezing, shortness of breath and/or trouble breathing?	Other comments about cough or breathing

BOWEL MOVEMENTS (BM)

How many a day?	Appearance (e.g., hard, soft, floats, greasy, loose)	Bad smell?	Gas? (occasional, frequent or excessive)	Other (describe)

OTHER DIGESTIVE SYMPTOMS

Abdominal pain	Reflux	Appetite different from normal	Rectal prolapse	Other (describe)

OTHER COMMENTS

Appendix E: CF Foundation–Accredited Care Centers

Some people with cystic fibrosis (CF) live near only one CF Foundation–accredited care center. Others have a choice of several centers. Here are some questions to ask yourself about your CF care center.

- How well do you or your family work with the doctor and other center team members? Do you trust the CF care center team? Sometimes the fit may be better with one team than with another.
- How far away is the center? Will the location work out for you and your family if you or your child has to stay at the hospital for treatment?
- Is there a program for adults with CF?
- Does the center and hospital accept your insurance?
- Where is the hospital that people with CF are admitted?
- What services are available at the hospital?
- Are parents allowed to stay with their child? Is there a playroom? Is a teacher available at the hospital to help with schoolwork? Is there a child life specialist who can help your child cope with staying at the hospital?
- For adults with CF, can family members stay? Is there Internet access so you can continue to work or go to school?

To inspire action and strengthen partnerships, the CF Foundation provides care center data for key health measures for each of its accredited care centers at **www.cff.org**. This information is a tool for people with CF and their families to use to partner with their care center to improve CF care.

It may be helpful to visit each of the CF care centers near you to find out which will best fit your needs or the needs of your child and family. Remember, CF care teams at CF Foundation–accredited CF care centers have the most current information about treatments to keep you or your child in the best health possible.

The CF Foundation is available at **1-800-FIGHT CF** and **info@cff.org** to answer questions about your local CF care centers. Also see the additional care center–related resources in Appendix B.

Alabama

The Children's Hospital/UAB (Pediatric)
1600 7th Ave. South
Birmingham, AL 35233
(205) 638-9583
Hector H. Gutierrez, M.D.
Wynton Hoover, M.D.

University of Alabama at Birmingham (Adult)
The Kirklin Clinic
2000 6th Ave. South
Birmingham, AL 35205
(205) 975-3258
Veena Antony, M.D.
Kevin Leon, M.D.

Alaska

Providence Alaska Medical Center (Pediatric and Adult)
Pediatric Subspecialty Clinic
3300 Providence Dr., Suite 314
Anchorage, AK 99508
(907) 212-4824
Dion Roberts, M.D.
Elizabeth Galloway, M.D.

Arizona

Phoenix Children's Hospital (Pediatric and Adult)
1919 East Thomas Rd.
Phoenix, AZ 85016
(602) 933-0985
Pediatric: Peggy Radford, M.D.
Adult: Gerald D. Gong, M.D.

Tucson CF Center (Pediatric and Adult)
535 N. Wilmot Rd., Ste. 100
Tucson, AZ 85711
(520) 694-9988
Wayne J. Morgan, M.D., C.M.
Cori Daines, M.D.

Arkansas

Arkansas Children's Hospital (Pediatric)
800 Marshall St.
Little Rock, AR 72202
(501) 364-4000
John Carroll, M.D.
Gulnar Com, M.D.

University of Arkansas for Medical Sciences (Adult)
4301 W. Markham St., #555
Little Rock, AR 72205
(501) 603-1400
Paula Anderson, M.D.

California

Marin (Outreach CF Clinic)
1100 Larkspur Landing Circle, Suite 150
Larkspur, CA 94939
(415) 461-3498
Patty Sylvester

Miller Children's Hospital (Pediatric)
2801 Atlantic Ave.
Long Beach, CA 90806
(562) 933-8749
Eliezer Nussbaum, M.D.
Terry Chin, M.D., Ph.D.

Long Beach Memorial Medical Center (Adult)
2801 Atlantic Ave.
Ground Floor
Long Beach, CA 90806
(562) 933-2820
Jeffrey Riker, M.D.
Inderpal Randhawn, M.D.

Children's Hospital of Los Angeles (Pediatric)
4650 Sunset Blvd. MS# 83
Los Angeles, CA 90027
(323) 361-4545
Thomas Keens, M.D.

USC Keck School of Medicine (Adult)
1500 San Pablo St.
Los Angeles, CA 90033
(323) 442-8330
Adudpa Pursh Rao, M.D.

Kaiser Foundation Los Angeles Medical Center (Pediatric and Adult)
4700 Sunset Blvd., Module 3C
Los Angeles, CA 90027
(800) 954-8000
Muhammad Saeed, M.D.
Tina Chou, M.D.

Children's Hospital Central California (Pediatric)
9300 Valley Childrens Place
Madera, CA 93636
(559) 353-5550
Reddy Sudhakar, M.D., Ph.D.

Children's Hospital at Oakland (Pediatric)
747 52nd St., OPC-5409
Oakland, CA 94609
(510) 428-3314
Karen A. Hardy, M.D.

California Pacific Medical Center (Adult)
2351 Clay St., #501
San Francisco, CA 94115
(415) 923-3421
Ryan Dougherty, M.D.

Kaiser Foundation Oakland (Pediatric)
12th Floor, Pediatric Subspecialty
3505 Broadway
Oakland, CA 94619
(510) 752-6906
Gregory F. Shay, M.D.

Kaiser Foundation Oakland (Adult)
280 West MacArthur
Oakland, CA 94611
(510) 752-6906
Bryon Quick, M.D.

Children's Hospital of Orange County (Pediatric)
Pulmonary Clinic
455 South Main St.
Orange, CA 92868
(714) 532-7983
Bruce Nickerson, M.D.

Stanford University Medical Center (Pediatric)
Mary Johnson Ambulatory Care Center
730 Welch Rd.
Palo Alto, CA 94304
(650) 497-8841
Carlos E. Milla, M.D.

Stanford University (Adult)
300 Pasteur Dr.
Stanford, CA 94305
(650) 498-6840
Paul Mobabir, M.D.

Pleasanton (Outreach CF Clinic)
5820 Stoneridge Mall Rd.
Suite 210
Pleasanton, CA 95488
(925) 463-8970
Patty Sylvester

Kaiser Foundation Roseville (Pediatric)
Department of Pediatric Pulmonology
1600 Eureka Rd. - MOB II
Roseville, CA 95661
(916) 474-2267
Gregory F. Shay, M.D.

Sutter Medical Center (Pediatric)
1625 Stockton Blvd, Suite 112
Sacramento, CA 95816
(916) 262-9114
Bradley E. Chipps, M.D.

University of California at Davis Medical Center (Pediatric)
Glasrock Clinic Building
2521 Stockton Blvd
Sacramento, CA 95817
(916) 734-3112
Ruth McDonald, M.D.

This listing is current as of June 2012. For the most up-to-date list of care centers, visit the CF Foundation's website (www.cff.org) or call 1-800-FIGHT CF.

University of California at Davis Medical Center (Adult)
2825 J St., Suite 400
Sacramento, CA 95816
(916) 734-3738
Brian Morrissey, M.D.
Carroll Cross, M.D.

Kaiser Foundation South Sacramento (Adult)
MOB 2 Pulmonary Dept. 322
6600 Bruceville Rd.
Sacramento, CA 95823
(916) 688-2200
Bryon Quick, M.D.

Loma Linda University Medical Center (Pediatric)
2195 Club Center Dr., Suite G
San Bernardino, CA 92408
(909) 835-1808
Yvonne Fanous, M.D.
Henry Opsimos, M.D.

Rady Children's Hospital and Health Center at the University of California San Diego (Pediatric)
3020 Children's Way
San Diego, CA 92123
(858) 966-5846
Mark S. Pian, M.D.

University of California San Diego Medical Center-Thornton (Adult)
9300 Campus Point Dr., MC 7381
La Jolla, CA 92037
(858) 657-7073
Douglas J. Conrad, M.D.

Naval Medical Center San Diego–FOR MILITARY PERSONNEL ONLY (Pediatric and Adult)
34800 Bob Wilson Dr.
Department of Pediatrics
San Diego, CA 92134
(619) 532-6896
Henry Wojtczak, M.D.

The Regents of the University of California at San Francisco (Pediatric)
UCSF Pediatric Specialties Clinic
400 Parnassus Ave., 2nd Floor
San Francisco, CA 94143
(415) 353-7337
Dennis W. Nielson, M.D., Ph.D.

University of California at San Francisco (Adult)
UCSF Faculty Chest Practice
UCSF Ambulatory Care Center
400 Parnassus Ave., 5th Floor
San Francisco, CA 94143
(415) 353-2961
Mary Ellen Kleinhenz, M.D.

California Pacific Medical Center (Outreach CF Clinic)
3700 California St., B554
San Francisco, CA 94115
(510) 428-3305
Patty Sylvester

Kaiser Permanente Foundation (Pediatric)
Pediatrics, MOB 190
710 Lawrence Expressway
Santa Clara, CA 95051
(510) 752-7098
Gregory F. Shay, M.D.

Kaiser Foundation Santa Clara (Adult)
Pulmonary Dept 282
710 Lawrence Expressway
Santa Clara, CA 95051
(408) 851-1161
Bryon Quick, M.D.

Tahoe Forest Multi-Specialty Clinic (Outreach CF Clinic)
10956 Donner Pass Rd., Suite 210
Truckee, CA 96161
(530) 587-3523
Estella Iniguez

Pediatric Diagnostic Center (Pediatric and Adult)
3160 Loma Vista Rd.
Ventura, CA 93003
(805) 652-6255
Chris Landon, M.D.

Walnut Creek (Outreach CF Clinic)
2401 Shadelands Dr.
Walnut Creek, CA 94598
(925) 979-4000
Patty Sylvester

Colorado

The Children's Hospital Colorado (Pediatric)
Pediatric Pulmonary Medicine
Cystic Fibrosis Program
Children's Hospital, Box B395
13123 East 16th Ave.
Aurora, CO 80045
(720) 777-6181
Scott Sagel, M.D., Ph.D.

National Jewish Medical and Research (Adult)
1400 Jackson St.
Denver, CO 80206
(303) 398-1178
Jerry A. Nick, M.D., B.S.

Children's Hospital Colorado Outpatient Specialty Care (Outreach CF Clinic)
4125 Briargate Parkway
Colorado Springs, CO 80920
(720) 777-5869
Melissa Burkey

Pediatric Partners of the Southwest
575 Rivergate Lane, Suite 109
Durango, CO 81301
720-777-5869
Melissa Burkey

Connecticut

Central Connecticut Cystic Fibrosis Center (Pediatric)
Pediatric Pulmonology
Connecticut Children's Medical Center
282 Washington St.
Hartford, CT 06106
(860) 545-9440
Craig D. Lapin, M.D.

Central Connecticut Cystic Fibrosis Center (Adult)
80 Seymour St.
Pulmonary Lab: Jefferson Building
Hartford, CT 06102
(860) 545-4644
James Pope, M.D.

Yale University School of Medicine (Pediatric)
Yale New Haven Children's Hospital
2nd Floor Subspecialty Center
20 York St.
New Haven, CT 06520
(203) 785-4081
Marie E. Egan, M.D.

Yale University School of Medicine (Adult)
Winchester Clinic
20 York St., 2nd Floor
New Haven, CT 06520
(203) 785-4198
Jonathan L. Koff, M.D.

Delaware

Alfred I. duPont Hospital for Children (Pediatric and Adult)
1600 Rockland Rd.
WIlmington, DE 19803
(302) 651-6400
Raj Padman, M.D.
Aaron Chidekel, M.D.

District of Columbia

Children's National Medical Center (Pediatric and Adult)
111 Michigan Ave., NW
Suite 1030
Washington, DC 20010
(202) 476-2128
Pediatric: Anastassios Koumbourlis, M.D., M.P.H.

Adult: Peter Levit, M.D.

This listing is current as of June 2012. For the most up-to-date list of care centers, visit the CF Foundation's website (www.cff.org) or call 1-800-FIGHT CF.

Florida

Children's Hospital of Southwest Florida/Children's Specialists Pediatric Pulmonology & Sleep Medicine (Pediatric)
7970 Summerlin Lakes Drive
Fort Myers, FL 33905
(239) 437-5500
Oscar A. Alea, M.D.

University of Florida (Pediatric)
Medical Plaza Pediatric Specialties Clinics
2000 SouthWest Archer Rd.
Gainesville, FL 32608
(352) 273-8381
Pamela M. Schuler, M.D.

University of Florida (Adult)
1600 SW Archer Rd., Suite 1901
POB 100383
Gainesville, FL 32610
(352) 273-8735
Mark Brantly, M.D. (Interim)

Joe DiMaggio Children's Hospital (Pediatric and Adult)
3341 Johnson St.
Hollywood, FL 33021
(954) 265-6333
Juan Martinez, M.D.

Nemours Children's Clinic–Jacksonville (Pediatric)
807 Children's Way
Jacksonville, FL 32207
(904) 697-3788
Bonnie B. Hudak, M.D.
David Schaeffer, M.D.

Pulmonary and Critical Care Associates of Jacksonville (Adult)
425 North Lee St.
Suite 202
Jacksonville, FL 32204
(904) 366-3738
Harish Bhaskar, M.D.

University of Miami (Pediatric and Adult)
Batchelor Children's Research Institute
1580 NW 10th Ave., 1st Floor
Miami, FL 33136
Pediatric: (305) 243-6162
Annabelle Quizon, M.D.
Andrew R.A. Colin, M.D.
Adult: (305) 243-6388
Matthias Salathe, M.D.

Miami Children's Hospital (Pediatric)
3200 SW 60 Court
Medical Arts Building, Suite 203
Miami, FL 33155
(305) 669-5864
Maria E. Franco, M.D.

Arnold Palmer Hospital Specialty Practice (Pediatric)
83 West Columbia St.
Orlando, FL 32806
(321) 841-6317
Mark Weatherly, M.D.

Nemours Children's Clinic–Orlando (Pediatric)
1717 South Orange Ave.
Orlando, FL 32806
(407) 650-7270
Floyd Livingston, M.D.

Central Florida Pulmonary Group (Adult)
326 North Mills Ave.
Orlando, FL 32803
(407) 841-1100
Francisco Calimano, M.D.
Daniel Layish, M.D.

Nemours Children's Clinic - Pensacola (Pediatric and Adult)
5153 N. 9th Ave.
Pensacola, FL 32504
(850) 505-4785
Barbara Stewart, M.D.

All Children's Specialty Care Clinic of Sarasota (Outreach CF Clinic)
5811 Rand Blvd.
Sarasota, FL 34238
(727) 767-3995
Stasia Lehmann, RN, BSN

All Children's Hospital (Pediatric)
601 5th St. South, Suite 708
St. Petersburg, FL 33701
(727) 767-3995
Magdalen Gondor, M.D.

Tampa General Hospital (Adult)
5 Tampa General Circle, Suite 300
Tampa, FL 33606
(813) 844-4634
Mark Rolfe, M.D.

University of South Florida (Pediatric)
13101 Bruce B. Downs Blvd.
Tampa, FL 33612
(813) 259-8767
Bruce M. Schnapf, D.O.

Georgia

Georgia Regents University (Pediatric)
1120 15th St.
Augusta, GA 30912
(706) 721-2635
Katie McKie, M.D.

Georgia Regents University (Adult)
1120 15th St.
Augusta, GA 30912
(706) 721-4658
Caralee Forseen, M.D.

Emory University (Pediatric)
2015 Uppergate Dr.
Atlanta, GA 30322
(404) 727-5728
Michael Schechter, M.D.

Emory University (Adult)
1365 Clifton Rd. N.E.
4th Floor, Bldg A #4325
Atlanta, GA 30322
(404) 778-7928
Seth Walker, M.D.

Children's Healthcare of Atlanta at Scottish Rite (Pediatric)
1100 Lake Hearn Dr.
Suite 450
Atlanta, GA 30342
(404) 785-2898
Kevin Kirchner, M.D.

Hawaii

Tripler Army Medical Center– FOR MILITARY PERSONNEL ONLY (Pediatric and Adult)
1 Jarrett White Rd.
Honolulu, HI 96859
(808) 433-5907
Jane E. Gross, M.D., Ph.D.

Idaho

Saint Luke's Cystic Fibrosis Center of Idaho (Pediatric and Adult)
100 E. Idaho St.
Suite 200
Boise, ID 83712
(208) 381-7092
Henry R. Thompson, M.D.

Illinois

Ann & Robert H. Lurie Children's Hospital of Chicago (Pediatric)
225 E. Chicago Ave.
Chicago, IL 60611
(312) 227-6730
Susanna A. McColley, M.D.
Adrienne L. Prestridge, M.D.

Northwestern University (Adult)
Suite 14-002B
676 N. St. Clair
Chicago, IL 60611
(773) 880-4382
Manu Jain, M.D.

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Rush University Medical Center (Pediatric)
Pediatric Subspeciality Clinic
1725 W. Harrison St., POB 710
Chicago, IL 60612
(312) 942-3034
Girish Sharma, M.D.
John Lloyd-Still, M.D.

Rush University Medical Center (Adult)
1725 W. Harrison, POB 054
Chicago, IL 60612
(312) 942-6744
Robert A. Balk, M.D.

University of Chicago (Pediatric)
5721 S. Maryland Ave.
Comer 2, 4th Floor
Chicago, IL 60637
(773) 702-6178
Lucille A. Lester, M.D.

University of Chicago (Adult)
5841 S. Maryland Ave.
DCAM, 4th Floor
Chicago, IL 60637
(773) 702-9660
Edward Naureckas, M.D.

Loyola University Medical Center (Pediatric and Adult)
2160 S. First Ave.
Maywood, IL 60153
(708) 327-9134
Sean Forsythe, M.D.

Advocate Lutheran General Children's Hospital CF Center (Pediatric)
1675 Dempster St.
2nd Floor
Park Ridge, IL 60068
(847) 318-9330
Gabriel Aljadeff, M.D.

Lutheran General Hospital (Adult)
North Suburban Pulmonary Specialists
8780 W. Golf Rd., Suite 102
Niles, IL 60714
(847) 759-4770
Arvey M. Stone, M.D.

Advocate Hope Children's Hospital (Pediatric)
4440 West 95th St.
Oak Lawn, IL 60453
(708) 684-5810
Javeed Akhter, M.D.

Saint Francis Medical Center (Pediatric and Adult)
320 East Armstrong Ave.
Peoria, IL 61603
(309) 624-6565
Pediatric: Jalayne M. Lapke, M.D., F.A.A.P.

Adult: W. Anthony Sauder, M.D.

Southern Illinois University School of Medicine (Pediatric and Adult)
751 North Rutledge St.
Room 0300
Springfield, IL 62702
(217) 545-5864
Mark Johnson, M.D.
Joseph Henkle, M.D.

Indiana

Deaconess Hospital (Outreach CF Clinic)
600 Mary St.
Evansville, IN 47710
(812) 858-3131
Sara King

Lutheran Children's Hospital (Pediatric and Adult)
7950 W. Jefferson Blvd.
Primary Life Services
Fort Wayne, IN 46804
(260) 435-7123
Pushpom Z. James, M.D.

Riley Hospital for Children Indiana University Medical Center (Pediatric)
Riley Hospital for Children
702 Barnhill Drive, ROC 2nd Floor
Indianapolis, IN 46202
(317) 274-7208
Michelle S. Howenstine, M.D.

Indiana University (Adult)
550 N. University Blvd.
Indianapolis, IN 46202
(317) 948-8660
Michael Ober, M.D.

Saint Joseph Regional Medical Center (Pediatric and Adult)
Pediatric Specialty Clinics
611 East Douglas Rd.
Suite 405
Mishawaka, IN 46545-1468
574-335-6240
James B. Harris, III, M.D.

Iowa

Mary Greeley Hospital–McFarland Clinic (Pediatric and Adult)
1215 Duff Ave.
Ames, IA 50010
(515) 239-4482
Edward. Nassif, M.D.

Blank Children's Health Center (Pediatric)
1212 Pleasant St. Ste. 204
Des Moines, IA 50309
(515) 241-6548
Ricardo Flores, M.D.

University of Iowa (Pediatric)
Pediatric Dept., UIHC
200 Hawkins Drive
Iowa City, IA 52242
(319) 356-2229
Miles Weinberger, M.D.
Richard Ahrens, M.D.

University of Iowa (Adult)
Medical Subspecialty Clinics UIHC
Department of Internal Medicine
200 Hawkins Drive
Iowa City, IA 52242
(319) 356-8133
Douglas Hornick, M.D.

Kansas

University of Kansas Medical Center (Pediatric)
3901 Rainbow Blvd
MS 4004
Kansas City, KS 66160
(913) 588-6364
Mitzi Scotten, M.D.

University of Kansas Medical Center (Adult)
Suite A Bell Hospital
Mailstop 3007
3901 Rainbow Blvd.
Kansas City, KS 66160
(913) 588-1227
Joel Mermis, M.D.
Steven Stites, M.D.

Via Christi–Saint Francis Campus (Pediatric and Adult)
707 N. Emporia
Wichita, KS 67214
(316) 858-3463
Pediatric: Natalie Sollo, M.D.

Adult: Janel Harting, M.D.

Kentucky

University of Kentucky (Pediatric)
Kentucky Clinic - 2nd Floor - Wing C
740 South Limestone St.
Lexington, KY 40536
(859) 323-6211
Jamshed F. Kanga, M.D.

University of Kentucky (Adult)
Kentucky Clinic - 5th Floor - Wing D
740 South Limestone St.
Lexington, KY 40536
(859) 323-9555
Michael I. Anstead, M.D.

University of Louisville (Pediatric)
234 E. Gray St.
Suite 270
Louisville, KY 40202
(502) 629-8830
Nemr S. Eid, M.D.

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University of Louisville (Adult)
401 E. Chestnut St., Suite 310
Louisville, KY 40202
(502) 813-6500
Rodney J. Folz, M.D., Ph.D.

Louisiana

Tulane University (Pediatric and Adult)
1415 Tulane Ave.
New Orleans, LA 70112
(504) 988-5800
Pediatric: Scott H. Davis, M.D.
Adult: Dean B. Ellithorpe, M.D.

Louisiana State University Health Sciences Center (Pediatric and Adult)
Department of Pediatric Pulmonary
1501 Kings Highway
Shreveport, LA 71103
(318) 675-6094
Kimberly L. Jones, M.D.

Maine

Eastern Maine Medical Center (Pediatric and Adult)
417 State St., Suite 305
Bangor, ME 04401
(207) 973-4051
Thomas Lever, M.D.

Maine Medical Partners Pediatric Specialty Care (Pediatric)
887 Congress St., Suite 320
Portland, ME 04102
(207) 662-5522
Anne Marie Cairns, D.O.

Maine Medical Center (Adult)
Chest Medicine Associates
100 Foden Rd.
South Portland, ME 04106
(207) 828-1122
Jonathan Zuckerman, M.D.

Maryland

Johns Hopkins University (Pediatric)
David Rubenstein Child Health Building
200 North Wolfe St., Lower Level
Baltimore, MD 21287
(410) 955-2795
Peter J. Mogayzel, Jr., M.D., Ph.D.
Pamela L. Zeitlin, M.D., Ph.D.

Johns Hopkins University (Adult)
601 N. Caroline St.
Baltimore, MD 21287
(410) 502-7044
Michael P. Boyle, M.D., F.C.C.P.

National Institutes of Health (Adult)
NIH Clinical Center
ACRF 9
10 Center Drive
Bethesda, MD 20892
(301) 496-6821
Milica S. Chernick, M.D.
James E. Balow, M.D.

National Naval Medical Center–FOR MILITARY PERSONNEL ONLY (Pediatric and Adult)
Pulmonology Clinic
8901 Rockville Pike
Bethesda, MD 20889
(301) 295-4959
Andrew J. Lipton, M.D., M.P.H., T.M.

Massachusetts

Children's Hospital Boston (Pediatric)
300 Longwood Ave.
Mailstop 208
Boston, MA 02115
(617) 355-1900
Henry L. Dorkin, M.D.

Bringham and Women's Hospital (Adult)
74 Francis St.
Center for Chest Diseases
Boston, MA 02115
(617) 355-1900
Ahmet Uluer, D.O.

Massachusetts General Hospital (Pediatric)
Joey O'Donnell Cystic Fibrosis Center
275 Cambridge St., Suite 530
Boston, MA 02114
(617) 726-8707
Samuel Moskowitz, M.D.

Massachusetts General Hospital (Adult)
55 Fruit St. Cox 2
Boston, MA 02114
(617) 724-0520
Leonard Sicilian, M.D.

Tufts Medical Center (Pediatric and Adult)
Floating Hospital for Children
2nd Floor
800 Washington St.
Boston, MA 02111
(617) 636-7917
William F. Yee, M.D.

Baystate Medical Center (Pediatric and Adult)
3300 Main St., Suite 4B
Springfield, MA 01199
(413) 794-0815
Robert Gerstle, M.D.

UMass Memorial Health Care–Pediatric Pulmonary, Asthma and CF Center (Pediatric)
Dept. of Pediatrics, S5-860
55 Lake Ave.
Worcester, MA 01655
(508) 856-4155
Brian P. O'Sullivan, M.D.

UMass Memorial Health Care (Adult)
Benedict Building, University Campus
55 Lake Ave., North
Worcester, MA 01655
(508) 856-4155
Oren Schaeffer, M.D.

Michigan

University of Michigan Health System (Pediatric and Adult)
1500 E. Medical Center Dr.
Taubman Center, Floor 1
Reception D
Ann Arbor, MI 48109
Pediatric: (734) 764-4123
Samya Nasr, M.D.
Adult: (734) 647-9342
Richard H. Simon, M.D.

Children's Hospital of Michigan (Pediatric)
3901 Beaubien Blvd.
Detroit, MI 48201
(313) 745-5541
Ibrahim Abdulhamid, M.D.

Wayne State University Harper University Hospital (Adult)
4201 St. Antoine, Suite 4C
Detroit, MI 48201
(313) 745-9151
Dana G. Kissner, M.D.
Ayman Soubani, M.D.

Hurley Children's Clinic at Mott Children's Health Center Outreach Program (Outreach CF Clinic)
One Hurley Plaza
Flint, MI 48503
(810) 257-9344
Cem Demirci, M.D.

Helen DeVos Women and Children's Center (Pediatric)
330 Barclay Avenue, NE
Suite 200
Grand Rapids, MI 49503
(616) 267-2200
John Schuen, M.D.

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Helen DeVos Women and Children's Center (Adult)
Spectrum Health, Butterworth Hospital
100 Michigan St.
Grand Rapids, MI 49503
(616) 267-8244
Stephen Fitch, M.D.

Western Michigan University School of Medicine Clinics (Pediatric and Adult)
1000 Oakland Drive
Kalamazoo, MI 49008
(269) 337-6433
Douglas Homnick, M.D., M.P.H.

Michigan State University CF Center – Lansing (Pediatric and Adult)
1200 E. Michigan Ave., Suite 145
Lansing, MI 48912
(517) 364-5440
Myrtha Gregoire-Bottex, M.D.

Minnesota

Children's Hospitals and Clinics (Pediatric)
2530 Chicago Ave. South
Suite 400
Minneapolis, MN 55404
(612) 813-3330
John McNamara, M.D.

University of Minnesota (Pediatric)
Pediatric Specialty Clinic
516 Delaware St. SE, 4th Floor
Minneapolis, MN 55455
(612) 625-5995
Warren Regelmann, M.D.

University of Minnesota (Adult)
516 Delaware St. SE, 2nd Floor
Minneapolis, MN 55455
(612) 625-5995
Jordan Dunitz, M.D.

Mississippi

University of Mississippi Medical Center (Pediatric and Adult)
2500 North State St.
Jackson, MS 39216
Pediatric: (601) 984-5205
Joseph Marc Majure, M.D.

Adult: (601) 815-1145
Nauman Chaudary, M.D.

Missouri

Children's Hospital University of Missouri Health Sciences Center (Pediatric and Adult)
1101 Hospital Dr.
Columbia, MO 65212
Pediatric: (573) 882-6978
James Acton, M.D.

Adult: (573) 882-6978
Melissa Kouba, M.D.

The Children's Mercy Hospital–University of Missouri at Kansas City (Pediatric and Adult)
Pulmonology and Cystic Fibrosis Clinics
Room 1740.00, 1st Floor
Out-Patient Care Center
2401 Gillham Rd.
Kansas City, MO 64108
(816) 983-6490
Philip Black, M.D.

Freemont Medical Building (Outreach CF Clinic)
1965 S. Freemont St., Suite 220
Freemont Medical Building
Springfield, MO 65807
(573) 882-6978
James Acton, M.D.

Cox South Medical Center (Outreach CF Clinic)
3443 S. National Ave.
Springfield, MO 65807
(417) 269-4856
John Carlile

St. Louis Children's Hospital Washington University School of Medicine (Pediatric)
One Children's Place
Ambulatory Clinic, Suite C
St. Louis, MO 63110
(314) 454-2694
Peter Michelson, M.D., M.S.

Washington University School of Medicine (Adult)
4921 Parkview Place, Suite 8B
St. Louis, MO 63110
(314) 454-8640
Daniel Rosenbluth, M.D.

Cardinal Glennon Children's Medical Center St. Louis University (Pediatric)
1465 S. Grand Blvd.
St. Louis, MO 63104
(314) 268-4107
Blakeslee E. Noyes, B.A., M.D.
Gary Albers, M.D.

Saint Louis University Medical Center (Adult)
3660 Vista Ave., Suite 200
St. Louis, MO 63110
(314) 977-6190
Ravi P. Nayak, M.D.

Montana

Billings Clinic (Pediatric and Adult)
2800 Tenth Ave. North
Billings, MT 59101
(406) 238-5137
Jerimiah Lysinger, M.D.

Nebraska

Lincoln–Outreach Clinic (Outreach CF Clinic)
445 South 86th St.
Lincoln, NE 68526
(402) 559-6275
Carla Pospisal

North Platte CF Clinic (Outreach CF Clinic)
210 McNeil Lane
North Platte, NE 69101
(402) 559-6275
Carla Pospisal

Children's Hospital and Medical Center
8200 Dodge St.
Omaha, NE 68114
(402) 559-6275
John L. Colombo, M.D.

University of Nebraska Medical Center (Adult)
42nd & Emile
Omaha, NE 68198
(402) 559-9101
Peter J. Murphy, M.D.

Nevada

University of Nevada School of Medicine Children's Lung Specialists (Pediatric)
Cystic Fibrosis Center of Southern Nevada
University Medical Center of Southern Nevada
2231 West Charleston Blvd.
Las Vegas, NV 89102
(702) 598-4411
Craig T. Nakamura, M.D.

Adult Cystic Fibrosis Center (Adult)
University Medical Center
Total Life Care
2231 West Charleston Blvd.
Las Vegas, NV 89107
(702) 598-4411
Angelica Honsberg, M.D.

Renown Regional Medical Center Children's Hospital (Pediatric and Adult)
1155 Mill St.
Reno, NV 89502
(775) 982-5123
Sonia Budhecha, M.D.

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New Hampshire

Dartmouth Hitchcock Medical Center (Pediatric)
Children's Hospital at Dartmouth
One Medical Center Drive
Lebanon, NH 03756
(603) 653-9884
Pamela Hofley, M.D.
Margaret Guill, M.D.

Dartmouth Hitchcock Medical Center (Adult)
Adult CF Clinic–Desk 5C
One Medical Center Drive
Lebanon, NH 03756
(603) 653-9884
H. Worth Parker, M.D.

New Jersey

Saint Barnabas Medical Center (Pediatric and Adult)
200 South Orange Ave.
Suite 225, Pediatric Specialties
Livingston, NJ 07039
(973) 322-7600
Dorothy S. Bisberg, M.D.

Monmouth Medical Center (Pediatric)
279 Third Ave., Suite 604
Long Branch, NJ 07740
(732) 222-4474
Robert L. Zanni, M.D.

Monmouth Medical Center (Adult)
279 Third Ave., Suite 303
Long Branch, NJ 07740
(732) 222-4474
Doantrang Du, M.D.

Goryeb Children's Hospital of Atlantic Health System (Pediatric)
100 Madison Ave.
Morristown, NJ 07960
(973) 971-4142
Arthur B. Atlas, M.D.

Morristown Memorial Hospital (Adult)
101 Madison Ave.
Morristown, NJ 07960
(973) 971-4103
Stanley B. Fiel, M.D.
Frederic Scoopo, M.D.

Bristol-Myers Squibb Children's Hospital at Robert Wood Johnson University Hospital (Pediatric)
Department of Pediatrics
Pulmonary Medicine and Cystic Fibrosis Center
CHINJ, RM 2300
89 French St.
New Brunswick, NJ 08901
(732) 235-5210
Thomas F. Scanlin, M.D.

Bristol-Myers Squibb Children's Hospital at Robert Wood Johnson University Hospital (Adult)
Adult Pulmonary
125 Paterson St.
New Brunswick, NJ 08901
(732) 235-5210
Sabina Hussain, M.D.

St. Joseph's Children's Hospital (Pediatric and Adult)
DePaul Ambulatory Center
11 Getty Ave., Second Floor
Paterson, NJ 07503
(973) 754-2550
Roberto V. Nachajon, M.D.

New Mexico

University of New Mexico School of Medicine (Pediatric and Adult)
University Hospital
2211 Lomas Blvd. NE, ACC 3
Albuquerque, NM 87106
Pediatric: (505) 272-6633
Lea Davies, M.D.
Adult: (505) 272-4751
Theresa Heyne Kamp, M.D.

New York

Albany Medical College (Pediatric)

Pediatric Pulmonary & Cystic Fibrosis Center
47 New Scotland Ave.
Albany, NY 12208
(518) 262-6880
Paul G. Comber, M.D., Ph.D.

Albany Medical College (Adult)

Pulmonary & Critical Care Medicine
47 New Scotland Ave.
Albany, NY 12208
(518) 262-5196
Jonathan M. Rosen, M.D.

Good Samaritan Hospital (Pediatric)

655 Deer Park Ave.
Babylon, NY 11702
(631) 321-2100
Louis Guida, Jr., M.D.

Women and Children's Hospital of Buffalo (Pediatric)

218 Bryant St.
Buffalo, NY 14222
(716) 878-7524
Drucy Borowitz, M.D.

Women and Children's Hospital of Buffalo (Adult)

219 Bryant St.
Buffalo, NY 14222
(716) 878-7524
Carla Frederick, M.D.

University Medical Center at Stony Brook (Pediatric)

37 Research Way
East Setauket, NY 11733
(631) 444-5437
Catherine Kier, M.D.

The Steven & Alexandra Cohen Children's Medical Center of New York (Pediatric)

Pediatric Cystic Fibrosis Center
865 Northern Blvd., Suite 103
Great Neck, NY 11021
(516) 622-5280
Joan K. DeCelie-Germana, M.D.

The Children's Hospital at Westchester Medical Center/ New York Medical College (Pediatric and Adult)

19 Bradhurst Ave., Suite 1400
Hawthorne, NY 10532
(914) 493-7585
Pediatric: Allen J. Dozor, M.D.

Adult: Caren Behar, M.D.

Beth Israel Medical Center Cystic Fibrosis Center (Pediatric and Adult)

1st Ave. & 16th St.
Bernstein Building, 7th Floor
New York, NY 10003
(212) 420-4100
Pediatric: Maria N. Berdella, M.D.

Adult: Patricia A. Walker, M.D.

Children's Hospital of New York Columbia University (Pediatric)

Children's Lung and Cystic Fibrosis Center
3959 Broadway, 7 Central
New York, NY 10032
(212) 305-5122
Meyer Kattan, M.D.

Columbia University Medical Center (Adult)

Gunnar Esiason Adult Cystic Fibrosis and Lung Program
622 West 168th St., PH 859
New York, NY 10032
(212) 305-0290
Emily DiMango, M.D.

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Mount Sinai School of Medicine (Pediatric)
Pediatric Pulmonary Division
Box 1202B
One Gustave L. Levy Place
New York, NY 10029
(212) 241-7788
Richard. J. Bonforte, M.D.
Andrew Ting, M.D.

New York University Medical Center (Pediatric and Adult)
NYU Fink Ambulatory Care Center
160 East 32nd Street
L-3 Medical
New York, NY 10016
(212) 263-5940
Robert Giusti, M.D.

University of Rochester Medical Center Strong Memorial Hospital (Pediatric)
601 Elmwood Ave., Box 667
Rochester, NY 14642
(585) 275-2464
Karen Z. Voter, M.D.
Clement L. Ren, M.D.

University of Rochester Medical Center Strong Memorial Hospital (Adult)
913 Culver Rd.
Rochester, NY 14609
(585) 654-5432
Robert Horowitz, M.D.

SUNY Upstate Medical University (Pediatric and Adult)
750 E. Adams St.
Syracuse, NY 13210
(315) 464-6323
Pediatric: Ran D. Anbar, M.D.
Adult: James Sexton, M.D.

Samaritan Medical Center (Pediatric and Adult)
830 Washington St.
Watertown, NY 13601
(315) 786-0254
Melynne Youngblood, M.D.

North Carolina

Mission Children's Clinic (Pediatric)
11 Vanderbilt Park Drive
Asheville, NC 28803
(828) 213-1740
Bruce K. Bacot, M.D.

Asthma & Allergy Specialists, PA (Pediatric)
8045 Providence Rd., Suite 300
Charlotte, NC 27277
(704) 341-9600
William Ashe, M.D.
Hugh R. Black, M.D.

University of North Carolina at Chapel Hill (Pediatric)
NC Children's Hospital
UNC Chapel Hill
101 Manning Drive
Chapel Hill, NC 27514
(919) 966-1401
Margaret W. Leigh, M.D.
George Retsch-Bogart, M.D.

University of North Carolina at Chapel Hill (Adult)
Adult Cystic Fibrosis Clinic
UNC Pulmonary Clinic
3400 Ambulatory Care Clinic
Mason Farm Rd., CB#7705
Chapel Hill, NC 27599
(919) 966-6838
Michael R. Knowles, M.D.
James R. Yankaskas, M.D.

Duke University Medical Center (Pediatric)
2301 Erwin Rd.
Durham, NC 27710
(919) 684-3364
Judith Voynow, M.D.
Thomas Miles Murphy, M.D.

Duke University Medical Center (Adult)
3116 North Duke St.
Durham, NC 27704
(919) 668-7360
Peter S. Kussin, M.D.

Wake Forest University Baptist Medical Center (Pediatric and Adult)
Medical Center Blvd.
Winston Salem, NC 27157
Pediatric: (336) 713-4500
Karl H. Karlson, Jr., M.D.

Adult: (336) 716-4843
Victor Ortega, M.D.

North Dakota

St. Alexis Heart and Lung Clinic (Pediatric and Adult)
310 N. 10th St.
Bismarck, ND 58501
(701) 530-7500
Pediatric: James A. Hughes, M.D.

Adult: Carla Zacher, M.D., F.A.A.P.

MeritCare Medical Center (Pediatric)
801 North Broadway
Fargo, ND 58122
(701) 234-6600
Stephen Tinguely, M.D.

Ohio

Children's Hospital Medical Center of Akron (Pediatric and Adult)
Lewis H. Walker Cystic Fibrosis Center
1 Perkins Square
Akron, OH 44308
(330) 543-3249
Pediatric: Nathan C. Kraynack, M.D.

Adult: Titus Sheers, M.D.

Cincinnati Children's Hospital Medical Center (Pediatric)
3333 Burnet Ave.
Cincinnati, OH 45229
(513) 636-6627
Gary McPhail, M.D.

University of Cincinnati Medical Center (Adult)
University Hospital
234 Goodman Ave.
Cincinnati, OH 45219
(513) 475-8523
Patricia M. Joseph, M.D.

University Hospitals Case Medical Center (Pediatric and Adult)
Rainbow Babies and Children's Hospital
11100 Euclid Ave.
Cleveland, OH 44106
(216) 844-7700
Pediatric: Michael W. Konstan, M.D.

Adult: Steven D. Strausbaugh, M.D. F.C.C.P.

Nationwide Children's Hospital (Pediatric and Adult)
700 Children's Drive
Columbus, OH 43205
(614) 722-4766
Pediatric: Karen S. McCoy, M.D.

Adult: John Heintz, M.D., Alpa Patel, M.D.

The Children's Medical Center of Dayton/Wright State University School of Medicine (Pediatric and Adult)
Children's Medical Center
One Children's Plaza
Dayton, OH 45404
(937) 641-3440
Pediatric: Robert J. Fink, M.D.

Adult: Gary M. Onady, M.D., Ph.D.

Northwest Ohio Cystic Fibrosis Center (Pediatric and Adult)
CF Care Center
2121 Hughes Drive, Suite 640
Toledo, OH 43606
Pediatric: (419) 291-2207
Pierre A. Vauthy, M.D.

Adult: (419) 291-4626
Jeffrey Lewis, M.D.

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Oklahoma

Oklahoma Cystic Fibrosis Center (Pediatric and Adult)
Pediatric Pulmonary and Cystic Fibrosis Clinic
1200 Phillips Ave., OUCPB
Suite 14101
Oklahoma City, OK 73104
(405) 271-6390
Pediatric: James Royall, M.D.
Adult: Kellie Jones, M.D.

Oklahoma Cystic Fibrosis Center–Tulsa (Pediatric and Adult)
OU Physicians - Pediatrics
444 E. 41st St., 2nd floor
Tulsa, OK 74135
(918) 502-2000
Joseph Walter, M.D.

Oregon

Oregon Health Sciences University (Pediatric)
OHSU Pediatric Cystic Fibrosis Center
Dept. of Pediatrics
707 SW Gaines, L106
Portland, OR, 97239
(503) 494-8023
Michael Wall, M.D.

Oregon Health Sciences University (Adult)
Physicians Pavilion
3181 SW Sam Jackson Park Road
UHN 67
Adult Cystic Fibrosis Clinic
Suite 320
Portland, OR 97239
(503) 494-1620
Gopal Allada, M.D.

Kaiser Permanente Northwest Region (Pediatric and Adult)
3550 N. Interstate Ave.
Portland, OR 97227
(503) 813-2000
Richard. C. Cohen, M.D.

Pennsylvania

Lehigh Valley Hospital & Health Network (Pediatric and Adult)
Pediatric Specialty Center
2545 Schoenersville Rd.
3rd Floor
Bethlehem, PA 18017
(610) 402-3844
Robert W. Miller, M.D.
Dharmesh Suratwala, M.D., M.B.B.A.

Saint Luke's Hospital (Outreach CF Clinic)
Route 512 and Broadhead
Leigh Valley Industrial Park
Bethlehem, PA 18017
(610) 954-4975
Laurie Varlotta, M.D.

Geisinger Medical Center (Pediatric)
100 N. Academy Ave.
Danville, PA 17822
(888) 675-5437
Carlos Perez, M.D.

Hershey Medical Center Pennsylvania State University (Pediatric and Adult)
500 University Drive
Hershey, PA 17033
Pediatric: (717) 531-5412
Gavin R. Graff, M.D.
Adult: (717) 531-6525
Robert L. Vender, M.D.

Children's Hospital of Philadelphia University of Pennsylvania (Pediatric)
3615 Civic Center Blvd.
Philadelphia, PA, 19104
(215) 590-3749
Ronald Rubenstein, M.D., Ph.D.

University of Pennsylvania Hospital (Adult)
Perelman Center for Advanced Medicine
3400 Civic Center Blvd.
Philadelphia, PA, 19104
(215) 662-3202
Denis Hadjiliadis, M.D., M.H.S.

Drexel University College of Medicine, St. Christopher's Hospital for Children (Pediatric)
Erie at Front St., Suite 2215
Philadelphia, PA 19134
(215) 427-5183
Laurie Varlotta, M.D.

Drexel University of Medicine, Hahnemann University Hospital (Adult)
Drexel Adult CF Center
219 N. Broad. St., 9th Floor
Philadelphia, PA 19107
(215) 762-2688
Jeffrey Hoag, M.D., M.S., FCCP

Children's Hospital of Pittsburgh of UPMC–University of Pittsburgh (Pediatric)
4401 Penn Ave.
3rd Floor, Main Building
Pittsburgh, PA 15224
(412) 692-5630
David M. Orenstein, M.D.
Daniel Weiner, M.D.

University of Pittsburgh School of Medicine (Adult)
4th Floor, Falk Medical Building
Comprehensive Lung Center
3601 Fifth Ave.
Pittsburgh, PA 15213
(412) 648-6161
Joel H. Weinberg, M.D.
Joseph M. Pilewski, M.D.

Rhode Island

Brown University Medical School Rhode Island Hospital Cystic Fibrosis Center (Pediatric and Adult)
Hasbro Children's Hospital
Respiratory and Immunology Center
593 Eddy St., Hasbro Lower Level
Providence, RI 02903
(401) 444-6540
Pediatric: Karen Daigle, M.D.
Adult: Walter Donat, M.D.

South Carolina

Medical University of South Carolina (Pediatric)
MUSC Pediatric Pulmonology
135 Rutledge Ave., Suite 279
Charleston, SC 29425
(843) 876-0444
Isabel Virella-Lowell, M.D.

Medical University of South Carolina (Adult)
Pulmonology and Critical Care Medicine
96 Jonathan Lucas St., Suite 812 CSB
Charleston, SC 29425
(843) 792-0729
Patrick A. Flume, M.D.

University of South Carolina (Pediatric)
9 Medical Park, Suite 505
Columbia, SC 29203
(803) 434-2505
Daniel C. Brown, M.D.

Greenville Hospital System University Medical Group (Pediatric)
200 Patewood Dr.
Suite A300
GHS UMG Pediatric Pulmonology
Greenville, SC 29615
(864) 454-5530
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South Dakota

Sanford USD Medical Center (Pediatric)
Sanford Children's Specialty Clinic
1600 W. 22nd St.
Sioux Falls, SD 57117
(605) 312-1000
James Wallace, M.D.

Sanford USD Medical Center (Adult)
1100 South Euclid Ave.
Sioux Falls, SD 57117
(605) 312-1000
Susan Rohr, M.D.
David Thomas, M.D.

Tennessee

T.C. Thompson Children's Hospital (Pediatric)
910 Blackford St.
Chattanooga, TN 37403
(423) 778-6501
Joel C. Ledbetter, M.D.

East Tennessee Children's Hospital (Pediatric)
2100 W. Clinch Ave.
Medical Office Building
Suite 310
Knoxville, TN 37916
(865) 541-8830
John Rogers, M.D.
Eduardo Riff, M.D.

University of Tennessee Medical Center (Adult)
University Pulmonary and Critical Care
1940 Alcoa Hwy, Ste. E-210
Knoxville, TN 37920
(865) 305-5888
Bruce Ludwig, M.D.

University of Tennessee Medical Center (Pediatric)
Pediatric Pulmonary Medicine
Physician's Office Building
777 Washington Ave. Suite P110
Memphis, TN 38103
(901) 287-5222
Dennis Stokes, M.D.

University of Tennessee Adult CF Program (Adult)
880 Madison
5th Floor, Suite A&B
Memphis, TN 38104
(865) 305-5888
Luis Murillo, M.D.

Vanderbilt Children's Hospital Vanderbilt University Medical Center (Pediatric)
2200 Children's Way
11215 Doctors' Office Tower
Nashville, TN 37232
(615) 343-7617
Rebekah Brown, M.D.

Vanderbilt University Medical Center (Adult)
1301 Medical Center Drive
Suite B-817, TVC
Nashville, TN 37232
(615) 322-2386
Bonnie S. Slovis, M.D.

Texas

Dell Children's Medical Center of Central Texas (Pediatric)
4900 Mueller Blvd.
Austin, TX 78723
(512) 324-0137
Bennie McWilliams, M.D.
Allan Frank, M.D.

Dell Children's Medical Center of Central Texas/UTSW Austin (Adult)
3305 Northland Dr., Suite 512
(512) 324-0137
Jason Fullmer, M.D.

Children's Medical Center of Dallas/University of Texas Southwestern (Pediatric and Adult)
1935 Medical District Drive
Dallas, TX 75235
(214) 456-2361
Pediatric: Carolyn Cannon, M.D., Ph.D.

Adult: Raksha Jain, M.D.

Tri-Services Military Cystic Fibrosis Center–FOR MILITARY PERSONNEL ONLY
(Pediatric and Adult)
3851 Roger Brooke Drive
Fort Sam Houston, TX 78234
(210) 916-4927
Pediatric: John M. Palmer, M.D.

Adult: Catherine Shoff, M.D.

Cook Children's Medical Center (Pediatric and Adult)
901 Seventh Ave., Suite 420
Fort Worth TX 76104
(682) 885-6299
Pediatric: James C. Cunningham, M.D.
Nancy L. Dambro, M.D.

Adult: John Burk, M.D.
Steven Q. Davis, M.D., M.S., F.C.C.P.

Baylor College of Medicine/ Texas Children's Hospital (Pediatric)
Pulmonary Medicine Service
6701 Fannin St., Ste. 1040
Houston, TX 77030
(832) 822-2778
Christopher M. Oermann, M.D.
Peter Hiatt, M.D.

Baylor College of Medicine/ Baylor Clinic/The Methodist Hospital (Adult)
6620 Main, Ste. 11B.15
Houston, TX 77030
(713) 798-2400
Marcia Katz, M.D.

Texas Tech University Health Sciences Center (Pediatric and Adult)
Texas Tech Physicians Pediatrics
3601 4th St., Mail Stop 9903
Lubbock, TX 79430
806-743-7336
Adaobi Kanu, M.D.

Christus Santa Rosa Children's Hospital (Pediatric and Adult)
David Goldsbury Center for Children and Families
Cystic Fibrosis Clinic
Second Floor
333 North Santa Rosa
San Antonio, TX 78207
Pediatric: (210) 704-2338
Donna Beth Willey-Courand, M.D.

Adult: (210) 223-3010
Pedro Sepulveda, M.D.

Children's Hospital at Scott & White (Pediatric and Adult)
2401 South 31st St., Building 27
Temple, TX 76508
(254) 724-5504
Alma Chavez, M.D.

University of Texas Health Center at Tyler (Pediatric and Adult)
11937 US Highway 271
Tyler, TX 75708
(903) 877-5270
Rudolfo Amaro, M.D.

Utah

Intermountain Cystic Fibrosis Center University of Utah Health Sciences Center (Adult)
University Hospital and Clinics
50 North Medical Drive - Clinic 3
Salt Lake City, UT 84132
(801) 585-2804
Ted G. Liou, M.D.
Holly Carveth, M.D.

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Intermountain Cystic Fibrosis Center University of Utah Health Sciences Center (Pediatric)
Pulmonology Division
100 N. Mario Capecchi Drive
Salt Lake City, UT 84113
(801) 662-1765
Barbara Chatfield, M.D.

Vermont

Vermont Children's Hospital (Pediatric and Adult)
Children's Specialty Center
Fletcher Allen Health Care
111 Colchester Ave.
ACC 4th Floor, East Pavillion
Burlington, VT 05401
Pediatric: (802) 847-8600
Thomas Lahiri, M.D.

Adult: (802) 847-1158
Laurie A. Leclair, M.D.

Virginia

University of Virginia (Pediatric)
2270 Ivy Rd.
Charlottesville, VA 22903
(434) 924-2250
Deborah K. Froh, M.D.

University of Virginia (Adult)
1220 Lee St.
Charlottesville, VA 22908
(434) 924-5219
Cynthia Brown, M.D.
Veronica Indihar, M.D.

Pediatric Lung Center (Pediatric and Adult)
2730A Prosperity Ave.
Fairfax, VA 22031
(703) 289-1410
John Osborn, M.D.

Children's Hospital of the King's Daughters Eastern Virginia Medical School (Pediatric and Adult)
601 Children's Ln.
Norfolk, Va. 23507
(757) 668-7137
Pediatric: Cynthia A. Epstein, M.D.

Adult: Ignacio Ripoll, M.D.

Naval Medical Center, Portsmouth–FOR MILITARY PERSONNEL ONLY (Pediatric and Adult)
Cystic Fibrosis Center
Department of Pediatrics
620 John Paul Jones Circle
Portsmouth, VA 23708
(757) 953-2955
Lori Vanscoy, M.D.
Nam Ly, M.D.

Virginia Commonwealth University (Pediatric and Adult)
Nelson Clinic 1st Floor
403 N. 11th St.
Richmond, VA 23298
(804) 828-2982
H. Joel Schmidt, M.D.

Washington

Seattle Children's Hospital (Pediatric)
4800 Sand Point Way NE
M/SA-5921
Seattle, WA 98105
(206) 987-2024
Ronald L. Gibson, M.D., Ph.D.

University of Washington Medical Center (Adult)
Medicine Subspecialties Clinic
1959 NE Pacific St.
Seattle, WA 98195
(206) 598-4215
Moira L. Aitken, M.D.

Pediatric Pulmonary and CF Clinic (Pediatric and Adult)
105 W 8th Ave., Suite 660E
Spokane, WA 99204
(509) 474-6960
Michael M. McCarthy, M.D.

Mary Bridge Children's Health Center (Pediatric and Adult)
311 So. L St., M/S 311-01-0C
Tacoma, WA 98415
(253) 403-3131
Lawrence A. Larson, D.O.
David H. Ricker, M.D.

Madigan Army Medical Center–FOR MILITARY PERSONNEL ONLY (Pediatric and Adult)
9040 A Fitzsimmons Drive
Tacoma, WA 98431
(253) 968-1878
Donald Moffitt, M.D.

West Virginia

West Virginia University Charleston Division (Pediatric and Adult)
830 Pennsylvania Ave., Suite 104
Charleston, WV 25302
(304) 388-1552
Raheel Khan, M.D., F.A.A.P.
Kevin Mupin, M.D.

West Virginia University CF Center (Pediatric)
1 Stadium Dr., POC
Morgantown, WV 26507
(304) 598-4835
Kathryn S. Moffett, M.D.

Wisconsin

Saint Vincent's Hospital (Pediatric and Adult)
835 S. Van Buren St.
Green Bay, WI 54301
(920) 433-8508
Peter Holzwarth, M.D.

Gundersen Lutheran Medical Center (Pediatric and Adult)
1900 South Ave.
La Crosse, WI 54601
(608) 775-4143
Todd Mahr, M.D.

University of Wisconsin (Pediatric)
American Family Children's Hospital
1675 Highland Ave.
Madison, WI 53792
(608) 263-6420
Michael J. Rock, M.D.

University of Wisconsin (Adult)
600 Highland Ave.
Madison, WI 53792
(608) 263-7203
Guillermo A. doPico, M.D.
Keith Meyer, M.D.

Marshfield Clinic (Pediatric and Adult)
1000 North Oak Ave.
Marshfield, WI 54449
(715) 387-5251
Keith Pulvermacher, M.D.

Children's Hospital of Wisconsin Froedtert & Medical College of Wisconsin (Pediatric)
9000 W. Wisconsin Ave.
PO Box 1997, MS B620
Milwaukee, WI 53201
(414) 266-6730
Diana Quintero, M.D.

Froedtert & Medical College of Wisconsin (Adult)
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2PV- Pulmonary Clinic
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ADDING TOMORROWS

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