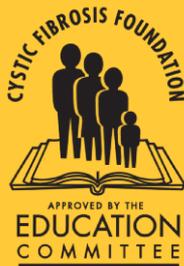


Beginning CF Care



For Parents of Children
with Cystic Fibrosis





PROGRAM DEVELOPED BY:

The CF Family Education Project
Baylor College of Medicine
and Texas Children's Hospital



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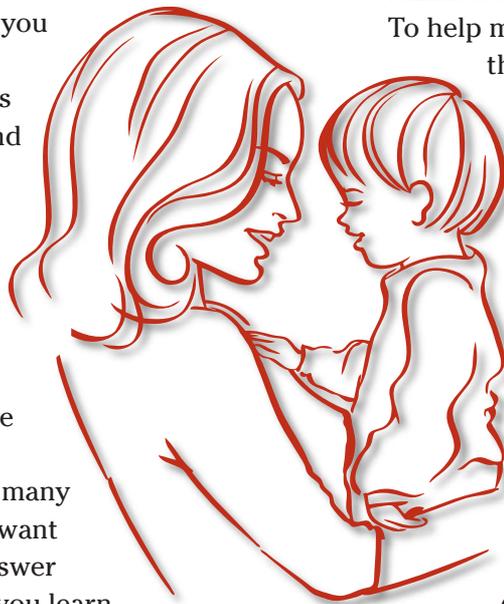


INTRODUCTION

When their child is **diagnosed*** with **cystic fibrosis*** (**CF***), most parents know very little about the illness. Many say that learning their child has CF is the hardest thing they've ever had to face. You probably have lots of questions and many different feelings. We have written this module to help you and your family begin to understand CF and manage your child's illness. The information in *Beginning CF Care* is especially written for parents of children younger than three who have just been diagnosed with CF.

In the beginning you may feel overwhelmed with how much there is to learn. With time and practice, you will master the skills necessary to care for your child. You will also become more confident. As you grow confident in your CF skills, you will become more actively involved in planning your child's care with the help of the CF health care team. The CF health care team wants you to be a partner in your child's care. You will work with your CF health care team to decide which skills in this module you want to learn first based on your child's medical problems and your family's needs.

Beginning CF Care focuses on information and skills that will help you get through the next few months. The CF health care team expects you to have many questions, and they want to be available to answer these questions. As you learn more about CF and CF care, you will work with the CF health care team on the other modules in the CF FAMILY EDUCATION PROGRAM (CF FEP) to become an expert CF care



manager. Over time, you can help your child to become a **CF manager*** too. We believe that this overview module is the best way to start gaining information about CF that you can use right away.

CF care has two goals:

- Keep the disease from getting worse.
- Help your child lead as normal a life as possible.

PROGRAM PHILOSOPHY

To help meet the overall goals of CF care, the CF FEP is based on the philosophy that taking care of CF be balanced with everything else in your family's and child's life. Having a child with CF can seem too much to handle. Parents of children with CF have so many tasks that they may lose the "big picture" with respect to the child's and family's life. This section provides the basic principles that guide much of the CF FEP.

Fostering Healthy Development in Your Child

Children with CF have medical **symptoms*** and treatment burdens beyond what most children experience. Because of this, some parents believe that they should focus on making the rest of the

*Glossary word. See [CF Words to Know Glossary](#).



child's life as carefree as possible.

The philosophy of the CF FEP differs from that idea. Helping your child develop normally and having the same expectations for your child that all parents do for their own children will lead to the best **quality of life*** for your child and family. Always being carefree is not helpful (and really not even possible!). Becoming **competent*** is better and, in the end, more rewarding for children. Notice how excited your toddler is when he or she learns a new task like catching a ball or using the potty. Children enjoy learning new things and you can expect your child to do most things that other children do.

Keeping a Balance Between Family Life and CF Care

Just as you cannot deny that CF is part of your life, you do not want CF to be your family's whole existence. Finding a balance for taking care of CF *and* living a normal life is an important goal of this program.

UNDERSTANDING CF

Cystic fibrosis is a **genetic*** disease. Genetic comes from the word "**genes**"* and means a disease, like CF, is **inherited.*** Genetic can mean that a trait or disease is inherited from one parent, but a child with CF inherits the disease from *both* parents. The child has the disease the moment he or she is **conceived.*** At conception the child receives a CF gene from the mother and a CF gene from the father. With two CF genes, the child will have CF. You can learn more about the genetics of CF from "THE GENETICS OF CYSTIC FIBROSIS" in Appendix 1. This may be helpful to you and anyone else in your family who is planning on having a child.



Cystic fibrosis is also a **chronic*** disease. This means your child will have it all of his or her life. We have no cure for CF yet, but we do

have treatments to control the symptoms and to slow the **progression*** of damage that CF causes.

The Diagnosis of CF

Parents usually find out that their child has CF in one of four ways:

- 1) A mother has genetic screening during pregnancy that leads to the child being tested
- 2) A newborn infant is tested because a state has a newborn screening program
- 3) A child is tested because of a family history of CF
- 4) A child is tested because of symptoms of CF

Your child may or may not have obvious symptoms at the time you learn the **diagnosis*** of CF.

If your child seems to have no obvious symptoms:

Your child's CF health care team will check to see if your child currently has any problems found in CF. The two most common problems are difficulty **digesting*** food and **lung infections.*** Sometimes problems can be so mild the symptoms are not noticed before the diagnosis. Your child may also have symptoms that were noticed, but were thought to be due to some other cause. For example, your child might have breathing problems that were thought to be caused by **allergies*** or frequent colds.

If your child is having problems related to CF, the CF health care team will decide what treatment your child needs. In addition, your CF health care team will want to begin CF care before problems or symptoms appear. The team will work with you to make a treatment plan, called a **CF action plan.*** They will also see your child regularly at the CF center to check on symptoms and treatments.

If your child has symptoms:

Your child's CF health care team will treat your child's obvious symptoms and check your child for other symptoms that may have gone unnoticed. For instance, if your child has lung infections, the doctors will also look to see if your child is having any problems with



digesting food. If the CF health care team finds that your child is having CF-related problems, the team will help you treat them.

Once your child's symptoms are under control, your child's CF health care team will work out what routine CF care your child needs every day. Using a treatment or CF action plan, you will learn what you should do when symptoms get worse (for example, what to do when your child has a lung infection).

The Most Common Problems in CF

The two most common problems in CF, partly caused by abnormal **mucus***, involve the **lungs*** (**respiratory system***) and the **digestive system***. The body makes mucus as a way to keep itself lubricated and clean (rather like motor oil in a car engine). In healthy people, the lungs and the **digestive tract*** both make mucus. In people with CF, the body makes mucus, but the mucus is different; it is much thicker and stickier. Abnormal mucus causes several problems.

The thick and sticky mucus in the lungs of people with CF is hard to **cough*** up. When mucus stays in the lungs, it makes an ideal place for **bacteria*** to grow. This is why most people with CF eventually have lung infections.

Most people with CF also have problems digesting food, and this is a big reason they may have problems with growing. The thick, sticky mucus blocks the **ducts*** from the **pancreas*** to the **small intestine***, so that the digestive **enzymes*** from the pancreas cannot get to the small intestine to help digest food. This is called **malabsorption*** or **pancreatic insufficiency***.

You will learn more about CF in the coming months. If you want more information right now, see "WHAT CAUSES PROBLEMS IN CYSTIC FIBROSIS?" in Appendix 2. But all you *have* to know right now is that children with CF usually have trouble digesting their food and are prone to lung infections.

CF TREATMENTS

Treatments that are now available can improve the symptoms of CF and slow down the progression of the disease. The CF health care team plans each child's care based on his or her medical problems. The CF health care team will teach you how to do these treatments.

Treatments for Digestive Problems

For the digestive problems of CF, most children take **pancreatic enzymes*** to help digest food, extra **vitamins*** and **minerals***, and most have a special high-calorie diet. You will manage the digestive problems of CF by learning how to:

- Feed your child
- Give enzymes
- Watch for malabsorption



You can see the parts of the digestive system in "CF FACTS — THE DIGESTIVE SYSTEM" in Appendix 3. You will be able to learn more in the CF FEP module *Managing Nutrition and Digestive Problems*.

Treatments for Lung Problems

Children take **antibiotics*** to fight lung infections. Also, **airway clearance*** and **breathing treatments*** (**inhaling*** medicine into the lungs) are used to help clear mucus from the lungs. You will manage lung problems by learning how to:

- Prevent lung infections
- Watch for **signs*** and symptoms of infection
- Manage infection
- Do airway clearance
- Give breathing treatments
- Give your child medicine
- Avoid tobacco smoke and other air pollution



You can see the parts of the lungs and respiratory system in "CF FACTS — THE RESPIRATORY SYSTEM" found in Appendix 4. You will be able to

*See CF Words to Know Glossary.



learn more in the CF FEP module *Managing Lung and Other Respiratory Problems*.

THE FUTURE

Nobody can predict how CF will affect your child in the future. In time, almost everyone with CF develops lung problems. But for each person, the age at which lung problems appear, how often they occur, and how quickly they progress is different.

Try not to compare your child's disease with that of other children who have CF. Some children who have CF have few or no symptoms for years. Others have many symptoms from the time they are babies. Symptoms may be mild or severe, and they may vary from child to child, even in the same family. Your child's symptoms may be different from one year to the next year.

Thirty years ago most children with CF died from the disease in childhood. That is not true any more. No one can say exactly how long any particular person with CF will live; however, more and more children with CF are living to be adults with happy, productive lives. Although CF is a life-shortening illness, more than half of people with CF in the United States have lived past their late thirties in recent years. Remember that the average life expectancy with CF will improve as **research*** helps find better ways of treating the disease.

Some parents say they worry that their child with CF may be fine one day and die the next. Actually, CF often progresses slowly. Lung damage is the most common cause of death, but it usually takes many years for the damage to progress and become life-threatening.

Thanks to research, the most serious lung problems can usually be delayed. Being able to benefit from the newest research requires people to be good disease managers and work with their CF health care team to learn how they can take advantage of the changes in CF care.

New Research and Treatments

There is no cure for CF at this time, but medical research continues to make steady progress. Researchers are always looking for new and better ways to help people prevent and slow down the progression of CF, such as new antibiotics to fight lung infections. Researchers are also developing new techniques to make CF care easier and more effective. For example, researchers are testing new equipment that will make breathing treatments faster and better: more medicine will be able to get directly into the lungs.

Researchers studying CF try to identify primary causes of CF, such as problems with how **chloride*** moves in and out of **cells*** and what genes are related to CF. New **therapies*** may try to focus on these basic causes. For example, ivacftor (brand name Kalydeco™) is a medicine that helps the **CFTR* protein*** work. It was first found to work in people who have a G551D **gene mutation***. (To learn more, see "WHAT CAUSES PROBLEMS IN CYSTIC FIBROSIS?" in Appendix 2.) **Gene therapy*** involves getting healthy genes to the cells to take over for the abnormal genes. The CF gene was found in 1989; this was the first step toward gene therapy. Gene therapy is still experimental and different approaches are being tested.

The CF Foundation is very committed to supporting and encouraging research to find a means to cure and control CF. You can learn more about CF research from your CF center and the CF Foundation Web site and newsletters (<http://www.cff.org>). You can also learn more about how you and your child may be a part of research studies.





Getting Started

MANAGING CYSTIC FIBROSIS

What you need to accomplish as a parent of a child with CF may seem overwhelming at first: You will need to get acquainted with your child's CF health care team, navigate the health care system, and cope with problems you have never heard of. At the same time, you need to be your child's parent. You will want to reach out for support from those around you and the CF health care team. What follows may help you sort out these challenges.

THE CF CENTER TEAM

CF centers have a CF health care team or group of professionals to work with each child and family. A variety of specialists may be on your child's CF health care team.

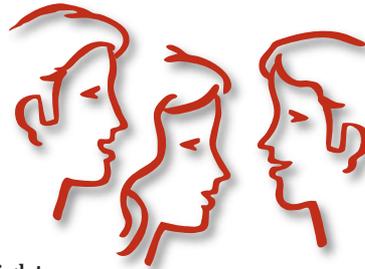
- Doctor who specializes in caring for children with CF
- Nurse who specializes in working with children with CF
- **Dietitian*** who helps you with your child's diet and enzymes
- **Social worker*** who may help you find the financial, physical, and emotional resources you need to cope with your child's needs
- Therapist who works with you on breathing treatments (he or she might be a **respiratory therapist*** or another member of the CF health care team)
- Therapist who will help you learn how to do airway clearance (he or she may be a **physical therapist,*** a respiratory therapist, or another CF health care team member)
- **Pharmacist*** who helps you understand your child's medicines and how to give them to your child

Not every child needs every specialist, and not every CF center has the same resources. Some CF centers may have other help on their teams, such as:

- **Genetic counselors***
- **Psychologists***
- **Child life specialists***
- School liaisons

- Teachers
- **Pediatric nurse practitioners*** or **physician assistants***

The center staff will tell you who is on your child's CF health care team. To record their names and phone numbers, use the worksheet "My Child's CF Health Care Team" in the back pocket of this module.



Your child's CF care team also includes you the parents, other family and friends who share in your child's care, and as your child gets older, he or she will become a more active part of the team as well. Each of you will be giving your child treatments and then watching to see how these treatments are working. Your active participation is vital to keep your child as healthy as possible.

You and Your Child's CF Health Care Team

Observing your child closely and watching for symptoms is another important part of your child's CF care. The professionals on the team are experts in their fields, but you are the expert on your child. You know your child better than anyone else; you see your child every day. You are the person most likely to notice small changes before they become big problems. You will most likely sense when "something just isn't right."

*See CF Words to Know Glossary.



To do your part as a member of your child's CF team, you will learn to become a good CF manager. You will learn skills called **Watch and Discover*** and **Think and Act*** to keep up with changes that could happen with this chronic disease. The module *Becoming a CF Manager* will help you learn these skills in more detail. This module, *Beginning CF Care*, will help you with the basics of CF care. Above all, you are your child's **advocate***. Don't be afraid to ask questions and pursue matters until you feel comfortable with the outcome.

Working with Your CF Center

You and your child will see the CF center team members for:

- CF checkups
- New or worsening symptoms



CF checkups are a routine part of CF care. The CF Foundation recommends that everyone with CF have quarterly visits (four times per year) to help watch for problems and do tests that

help make sure the disease is in good control. The module *Becoming a CF Manager* has more information about what to expect and how to prepare for CF checkups. Here are some of the things that the CF health care team does at checkups:

- Checks your child's progress and response to treatments
- Looks for subtle problems that you or your child's primary care provider may miss
- Helps you figure out your child's treatment or CF action plan and revises it based on any changes in your child's health and new research
- Makes sure that you (and later your child) know how to do CF treatments
- Helps your older child learn about CF, CF care, and accepting responsibility for his or her own care
- Helps your child enter adulthood as healthy as possible and with the skills needed to manage the disease and continue to stay healthy

Your child will need sick visits when he or she is having new or increased symptoms that are not getting better or going away with treatment. Your child's treatment plan may need adjusting, or there may be another problem to identify. Your child might be able to have a treatment plan prescribed for home or may need to be treated in the clinic or admitted to the hospital.

Write Down Questions for the Team

Writing down questions and what you observe about your child (*Watch and Discover*) before you contact the CF health care team is a good way to prepare for a visit or call to the CF center. Writing things down can help organize your thoughts. When you are worried or distracted by a squirming child, you may forget some of the concerns you have or things you have noticed that you want to discuss with the doctor or nurse.

- **Be Prepared to Answer Questions About Your Child's Symptoms** When you call about your child's symptoms, the team can help you better if you are prepared to answer certain questions. You will *Watch and Discover* at home for new or increased symptoms. When you notice changes in your child's signs or symptoms, the CF health care team would also like you to *Think and Act* even before you contact them. In some cases you will follow your child's CF action plan after noticing a change in symptoms and be ready to tell the team how it is working. You can learn more about how to *Watch and Discover* and *Think and Act* in the CF FEP module *Becoming a CF Manager*.
- **Find the Right Team Member to Ask** If you are not sure whom to contact about a particular question, ask your child's primary CF nurse. Your CF health care team members will help you figure out the best person for you to talk with.



- Make Sure You Understand the CF Health Care Team's Answers** You will need to convey what you learn at your CF clinic visits to other people who help care for your child. To do this, you will need to make sure you understand what is being said and what you are to do at home. If you don't understand an answer, ask again, or ask someone else to explain it. Some people find it helpful to take notes during visits. Repeat back to team members in your own words what they have told you. This is a great way to make sure you haven't misunderstood them and the team will appreciate your efforts to learn.



YOUR PRIMARY CARE PHYSICIAN AND WELL-CHILD CARE

Your **pediatrician*** or family **primary care physician*** is a part of your child's team. He or she focuses on well-child care and other health problems not related to CF. Your child should have all the well-child checkups and **immunizations*** that other children receive. Check with your primary care physician to find out what your child needs.

If your child is ill and you are not sure if the problem is related to CF, you can always call the CF center and the team will help you figure out whom you should take your child to see. You can also use the worksheet "Who to Call, Who to Visit" in the back pocket of this module to help guide you. You can fill out this form with the help of your CF health care team.

Some medical insurance providers will require that you get a referral from your primary care physician before your child is seen at the CF center. Call your primary care physician's office to arrange for a referral before your visit.

If your health care plan is restricting your child's access to the CF center, you need to be an advocate for your child. Ask to talk with the case manager for your insurance plan. Talk with the CF center social worker and your primary

care physician. You can read more about this issue in "WORKING WITH YOUR CF CENTER AND YOUR INSURANCE COMPANY" in Appendix 5.

WORKING WITH A PHARMACY

Your child will probably need several prescription medicines. Some parents use one pharmacy for regular prescriptions and another (one that is open late or makes deliveries) for emergencies. Your health care plan may also allow or require you to use a mail-order pharmacy for non-emergency medicines.

The CF Foundation operates a mail-order specialty pharmacy that many parents use because of its low cost and access to CF therapies. They also provide information about patient assistance programs. You may want to contact them at:

Cystic Fibrosis Services, Inc.
A Walgreens Alliance Pharmacy
6931 Arlington Rd., Suite 400
Bethesda, MD 20814
1-800-541-4959



The direct Web site is:

<http://www.walgreens.com/topic/pharmacy/cystic-fibrosis-services.jsp>.

THE CF FOUNDATION

The Cystic Fibrosis Foundation is a national organization headquartered in Bethesda, Maryland. Bethesda is near Washington, D.C., and is also the home of the National Institutes of Health (NIH). Along with the CF Foundation, NIH also sponsors research that is done in the United States.

The CF Foundation is committed to research and fundraising, and accredits about 275 CF centers across the United States. The CF Foundation checks on the performance of each CF center and requires that all centers have certain standards for care and resources to help people with CF and their families. The CF Foundation has a Web site that provides

*See CF Words to Know Glossary.



families with updates on CF research and education. The Web site shows where pediatric and adult CF centers are located and how to contact them. The CF Foundation also does regular Webcasts and other education programs for people with CF and their families. They have newsletters that are available free of charge.

The Web site address is <http://www.cff.org>. The CF Foundation's toll-free number is 1-800-FIGHT-CF (1-800-344-4823), or write:

Cystic Fibrosis Foundation
6931 Arlington Road, Suite 200
Bethesda, MD 20814



There are also local chapters of the CF Foundation in many cities. Local chapters help raise money for CF research, organize volunteers, and serve as an informal network for parents. Call the CF Foundation or check its Web site for the chapter nearest you.

MANAGING THE MEDICAL BILLS

Dealing with your child's diagnosis and the new demands of CF care are hard enough. Money is the last thing you want to worry about. Some things you can do are:

- Find a special place to keep CF-related bills
- Plan ahead and know your insurance plan
- Get in touch with people who can help you manage your medical bills:
 - Social worker at your CF center
 - Billing representative at the hospital (financial counselor)
 - Case manager at your insurance company

For more information, see "WORKING WITH YOUR CF CENTER AND YOUR INSURANCE COMPANY" in Appendix 5.

CF Center Social Worker

The social worker is an important member of your CF health care team. He or she has a lot of information about options and possible resources. You can ask your social worker general questions, such as "How am I going to afford this?" or "What am I eligible for?" Or you can ask specific questions, such as "Am I eligible for ...?"

Hospital Billing Representative/Financial Counselor

If your child has been in the hospital, you may have begun getting hospital bills. The hospital billing representative can be very helpful. Call this person if you have trouble:

- Keeping track of what has been paid and what has not
- Understanding your child's hospital bills
- Paying hospital bills—the billing representative can help you work out a payment plan with the hospital

Insurance Company Case Manager

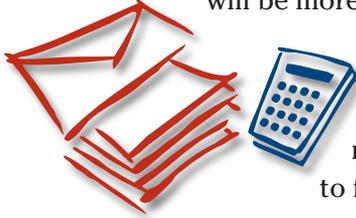
If you have medical insurance, call the company and find out if you have a case manager or if you can be assigned one. If you need help with this, ask your social worker.

A case manager can give your child's care more personal attention than the usual claims representatives. Your case manager can help you understand your policy and explain your insurance statements. He or she can also explain what your health care plan's policies are regarding CF care, and what the appeals process is if you want to challenge a decision. If you can form an alliance with your child's case manager, you may find this person a great help in the years ahead.



A Place for Bills

Find a place to keep your CF bills. Each time you get a bill, put it in that same spot as soon as you are through looking at it. That way, you will be more likely not to lose it or throw it away by mistake. Even if you're not ready to deal with bills right now, you will know where to find them when you are.



ADJUSTING TO CF

Many families say that they felt overwhelmed when their child was first diagnosed with CF. Few people are prepared to accept the news that their child has a chronic disease.

You may have any number of other feelings, such as:

- Fear for your child's health and how this disease will affect your family's lives
- Shock at finding that your child has a chronic disease
- Numbness
- Uncertainty about how your child will do in the future
- Sadness
- Anger or frustration
- Guilt
- Depression

In the period of time right after the diagnosis, any and all of these feelings are understandable and normal.

At the same time you are on this emotional roller coaster, you are expected to start learning how to do your child's CF care. Some people find it helpful to be learning what to do; others find it just one more burden. You will have to work through these feelings over time. Accepting CF as a diagnosis is the first important step in successfully learning how to manage the disease.

Here are some things that other parents have done to adjust to the diagnosis and management of the disease:

- Make CF treatments part of your daily routine
- Share responsibilities with your partner or family member
- Find people to talk to—both about your feelings and about how to manage CF
- Expect letdowns; do not expect to adjust quickly or perfectly
- Take some time for yourself and for your partner
- Enjoy your child—remember that CF is just one part of your child

Enjoy Your Baby

During the first year of life, the most important job for you and your baby is to get to know one another. At this age babies are learning love and trust. At the same time, you are getting to know your baby's smile, laugh, and unique personality. Even as you are learning about CF, take time to admire your baby. Cuddling and playing with your baby are just as important as learning about CF and CF care.

If your baby was diagnosed before having symptoms, you may be feeling very distressed—the baby you thought was well has now been diagnosed with a very serious disease. You may feel somewhat angry at the medical team for spoiling the happiness you have had bonding with your new child. These feelings will get better with time. Think about how you can continue to enjoy being a parent. Parents keep babies healthy and happy. Although your job just got harder, you can do it and the CF health care team is here to help you.

Problems from CF can make it harder to get to know your baby. He or she may have been sick or cranky. Some parents say that the hours spent in doctors' offices before their



child's illness was diagnosed left them worn out, angry, and frustrated. If your child has been having symptoms, once treatments start these symptoms will improve and your child will feel better. Your baby will have more energy and be less irritable. Enzymes will help your baby digest food and allow for normal growth. Your child will not feel hungry as often and will have fewer stomachaches and stools. Medicine and other treatments will help if your baby has a lung infection. Soon your child will be able to do more normal baby things, and you will have more energy to enjoy your baby.

Family and Friends

You will be talking to many people about your child. Some parents say it is difficult at first to talk about CF. They find that learning about CF and CF care makes it easier to explain to family members and friends about CF and its treatments. As you learn more about CF, you will also talk more easily with the members of your child's CF health care team.

You may hear a lot of well-meaning advice about what you should or should not do for your child with CF. People may say things like, "Why are you giving your child so much medicine? She doesn't look sick to me." Not everyone understands that your child needs daily care to stay as healthy as possible.



People also like to offer ideas on how to be a good parent. Some people may wonder why you are so cautious when they notice, for instance, that you keep your child away from children who are sick or have colds. They may say you are overprotective. Others may criticize you for the opposite reason. They may think that you aren't protective enough when you allow your child to go to day care or to play sports.

The advice and comments may be stressful. Talk with members of the CF health care team when you have concerns or are unsure about your child's care. They want to help you find ways to solve problems and learn new skills.

Even at this early stage, you already know more about CF than most people. As you gain experience managing CF every day, you will become even more of a CF expert. Don't be afraid to act on what you know. Soon you will feel more confident in making decisions that affect your child and his or her care. You can find more information on communicating with others about your child and CF care in the module

Becoming a CF Manager. For information on managing stress, also see the appendix in *Becoming a CF Manager*.

Child Cooperation and Acceptance

Throughout their lives, children with CF will need to do various treatments every day to delay or slow down the damage from CF problems. Helping children get into the habit of doing daily treatments when they are young is an important task for parents. Setting up a routine for treatments is much like setting up other basic routines in your child's life, such as brushing teeth and going to bed.



Doing the task every day, and at about the same time, is a good way to set up a routine. If a parent and child skip the task often, or especially if a child protests and a task is skipped, the child will learn that it is possible to get out of doing the task and it does not become routine. The effort you put into setting up a routine for CF care while your child is young will be well worth it in the long run. As part of a routine, CF care is more likely to be done every day and your child will be more likely to accept it and know it is expected.

Cystic fibrosis treatments are a burden that other children do not have. You can talk about the burden with your child, but do not dwell on the thought. A child who dwells on these tasks as burdens will not be using his or her energy wisely and will not feel competent to manage CF in the long run. Parents often need to manage their own reactions to CF care, as well as the reactions of their children, family, and friends, to avoid dwelling on the negative.

You can learn more about helping your child at different ages to be a good CF manager in the module *Working With Your Child*. You will also find tips on how to deal with problems in getting your child to cooperate with various treatments. You will learn more about what

to expect at different ages, such as when a child can learn to swallow pills. Managing CF is a journey you and your child will take together, with you as the guide.

Networking with Other CF Parents

Most parents do not worry about their two-year-old cooperating with **respiratory treatments*** because most parents do not have children with CF. Many parents find that it helps to talk with other parents of children with CF. These parents have gone through the same kinds of problems, are dealing with some of the same issues, and often have good ideas for solutions. Some CF centers have parent nights, support groups, or other ways for parents to get to know one another. Ask your CF social worker what is available.

Children with CF should not spend time together to limit the risk of passing certain bacteria among them. (Talk with your CF health care team to learn more.) Parents of children with CF, however, can get together. You may also find it helpful to connect with other parents through the computer. Your child may also be able to talk with other children with CF using the computer or phone.



*See CF Words to Know Glossary.



MANAGING YOUR CHILD'S NUTRITION

Most children with CF need more **calories*** than other children without CF to grow and stay healthy.

CF AND WEIGHT

A child with CF may have:

- Very slow weight gain
- No weight gain at all
- Weight loss, especially with **acute*** illness

The above problems are symptoms that a child is not getting enough calories. In a child with CF, these problems may be caused by:

- **Malabsorption:** The body has trouble digesting and **absorbing* nutrients*** from food and may not get enough calories even though the person is eating a normal amount.
- **Breathing problems:** The body uses more calories when it has to work harder to breathe; also, babies may find it hard to eat if they are having trouble breathing.
- **Infections:** The body needs more calories when it is fighting **germs*** that can often cause **infections*.**

Many people with CF have problems getting enough calories. To make sure they do get enough calories, most people with CF need to (1) eat a diet high in calories and nutrients, and (2) take enzymes to help digest and absorb their food better if they have malabsorption.

The CF health care team will help you learn how to Watch and Discover the symptoms and signs of growth and nutrition problems. They will help you Think and Act to make a plan for what to do to prevent and treat them. You can also learn more in the module *Managing Nutrition and Digestive Problems.*



WATCH AND DISCOVER

NUTRITION PROBLEMS

Growing and gaining weight is a major task in childhood. Babies and teens have the fastest rate of growth. For a child to get taller and gain weight, he or she needs to get enough calories and nutrients. As the child grows, the organs of the body, such as the brain and lungs, also continue to grow and develop. A problem with nutrition can stress the body and cause poor health as well as poor growth.

Every child will follow a pattern of growth over time that stays fairly consistent. This growth can be tracked on a chart. Doctors use **growth charts*** to look at how tall a child is getting and how well he or she is gaining weight over time. They also use charts to look at growth in the infant's head size. These charts show the normal range for children at each age in our population; what is most important is to see how each child compares to his or her own pattern over time. For example, if a child's weight has steadily increased over time, and then all of a sudden starts to drop, the doctor can tell that something is happening—the pattern has changed. The doctor can also estimate how tall a child may be expected to get compared to the height of his or her parents.





Every time you visit the CF center and your primary care doctor's office, your child will be measured. You should ask about your child's growth. Watch how your child is growing so you can Discover problems early. If you notice that your child has not changed clothing size for some time, you may suspect he or she is not growing well. If your child gets sick with a respiratory infection, you may notice some weight loss, and you'll know this is a time when your child will need more calories.



THINK AND ACT

MAINTAINING GOOD NUTRITION

Calorie Needs in CF

Calories are a way of measuring how much energy is in food. Nutrients, such as protein, vitamins, and minerals, are the parts of food that the body uses to live, grow, and repair itself.

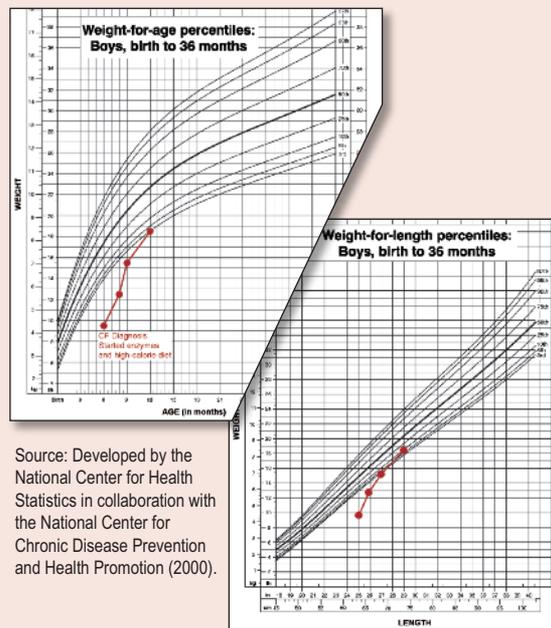
Children with CF need more calories than other children their age. Your child may need a special diet to meet these needs. Your baby's diet may include a special formula. Your toddler's diet may include nutritious, high-calorie snacks and/or **supplements*** along with regular meals.

Your dietitian can help you and your doctor decide if your child's calorie needs are being met. Your dietitian can also help you define a goal or target number of calories your child needs to grow and stay healthy. The dietitian may suggest a special formula or high-calorie dietary supplement to help reach your child's calorie goal.

WHAT YOU CAN DO

Using a Growth Chart

Ask for a copy of your child's growth charts. Have the CF dietitian or another team member show you how to keep track of your child's growth. You can bring the growth charts with you to future clinic visits and have the team help you add new measures as your child gets older. You can learn more about growth charts for children in the United States at the Centers for Disease Control and Prevention Web site (<http://www.cdc.gov/growthcharts/>).



Source: Developed by the National Center for Health Statistics in collaboration with the National Center for Chronic Disease Prevention and Health Promotion (2000).

Example Growth Charts: A newly diagnosed infant whose growth improved after he started eating a high-calorie diet and taking enzymes.

Using a Food Diary

If you are not sure whether your child is getting enough calories, you can keep a food diary. Write down everything your child eats and drinks in the diary, including the amount. (For example, record **6 ounces** of whole milk or **1/2 cup** of Jell-O brand chocolate snack pudding.) Your CF dietitian can tell you how to record the food and amount. You may keep the diary for a few days or a week. The dietitian can analyze this diary to give you an estimate of how many calories your child is getting. The dietitian can also look to see if your child is getting the right balance of different types of nutrients, such as protein or **calcium***. You can use "My Child's Food Diary," a worksheet in the back pocket of this module.

*See CF Words to Know Glossary.



Breast-Feeding

Breast-feeding has many benefits for you and your baby and you should continue to breast-feed as long as you are able to do so. Talk with the CF health care team. The CF dietitian and team will help you keep breast milk in your child's diet. They will help you weigh the benefits of breast-feeding and the benefits of formulas, which may make it easier to meet your child's special calorie needs.

Some infants who have CF grow well on breast milk alone. Babies who have malabsorption will need enzymes to help them digest breast milk. Some babies also need formula to get enough calories. If your baby needs formula, it is because he or she has CF, not because there is anything wrong with your breast milk.

Formula Feeding

If you are bottle-feeding your baby, the CF health care team may suggest you use a regular infant formula, but at a higher concentration (more powder), so each bottle has more calories. **This is NOT something you should do on your own.** If your child needs more calories, the team will show you how to prepare the formula. **Follow their instructions carefully.** Diluting a formula with too little water can cause as much damage as diluting a formula with too much water.

The team may also recommend a special type of infant formula to meet your child's dietary needs. These formulas contain the calories and nutrients your child needs in a form that is easier to digest. Some examples include:

Nutramigen® LIPIL®
Pregestimil® LIPIL®
Similac® Alimentum® Advance®

Babies who have malabsorption will need enzymes to digest most types of formula.

High-Calorie Food Additives

The CF health care team may suggest some products (called **high-calorie food additives***) that can be added to food to increase the number of calories. Some examples are:

- **Polycose** — a powder that can be added to food and drinks to add calories without changing the taste.
- **MCT oil** — a **fat*** solution that is easier for children with CF to digest than other fats.
- **Corn oil** — a liquid form of fat that can be used to increase calories.

When your child can eat solid foods, a CF health care team member will discuss with you whether your baby or toddler needs a high-calorie food additive. Increasing calories may be as simple as adding butter or cream to food.

High-Calorie Drinks

Many special ready-to-drink beverages are available to help meet your toddler's special calorie needs. Some examples are Pediasure®, Nutren Junior®, and Peptamen Junior.®

For older children, high-calorie supplements include Ensure® and Ensure Plus®, Equate® and Equate Plus®, Boost® and Boost® Plus, and SCANDISHAKE®. Some children can use breakfast drink powders (such as Carnation® Instant Breakfast®). You can also make your own high-calorie milk shakes. The dietitian or another member of the CF health care team can help you figure out what your child needs.



Vitamins and Minerals

Most people with CF do not get enough vitamins and minerals from the food they eat. For instance, they may have low levels of **fat-soluble*** vitamins A, D, E, and K or they may not get enough of certain minerals, such as **zinc***, **magnesium***, and calcium. Foods usually contain plenty of vitamins and



minerals, but a person with CF cannot always absorb enough of them. When this happens, the person needs to take extra vitamins and minerals or a vitamin-mineral product made especially for people with CF. The CF center usually checks blood levels of fat-soluble vitamins, magnesium, and calcium at least once a year and adjusts your child's **doses*** as needed.

Your child's doctor will prescribe the type and amounts of vitamins and minerals you should give your child. **Be sure to give only the amount prescribed by your CF health care team.** Too much of some vitamins can be harmful. ADEKs®, Vitamax®, and SourceCF® are some examples of vitamins made for people with CF. If you buy vitamins not made for children with CF, such as the chewable vitamins from the grocery store, your doctor may also need to prescribe doses of other vitamins. Look at the label on the vitamin supplement you use. Bring the label or a list of ingredients to your CF center and go over it with the CF dietitian.

Salt Intake

People with CF lose more salt in their sweat than do other people, which can cause a dangerous imbalance in the body's chemistry.

Breast milk, commercial baby foods, and infant formulas contain very little salt; therefore, you will need to add extra salt to your baby's diet. Talk with your CF health care team about how much extra salt to add.



As children get older, they usually begin to like more salty foods. Since there is enough salt in most foods, losing too much salt becomes less of a problem. During hot weather or with lots of activity, however, your child may need even more salt and fluids. Talk with your CF doctor or dietitian about how to add enough salt to your child's diet.

WHAT YOU CAN DO

Use the worksheets "What to Feed My Baby" or "What to Feed My Child" in the back pocket of this module and work with the dietitian or other member of the CF health care team on what to feed your infant or child. Many families prefer having information about enzymes and meals in one place. You will notice there are blanks to list the number of enzymes your child needs to take with each meal or snack. Most children with CF take enzymes with food. You will learn more about malabsorption and using enzymes in this module. You can photocopy the worksheet and share it with others who help feed your child.



MAKING IT WORK FOR YOU

Finding Special Formulas or Vitamin-Mineral Supplements

You can find many formulas at the grocery store or pharmacy. The same is true of vitamin and mineral supplements. If you can't find something the CF health care team has suggested, call and talk with the CF center dietitian or nurse. He or she may know who carries it. You can also talk to store managers or pharmacists to see if they can order it for you. You may also want to check with the CF Services Pharmacy for a supply (see earlier section on *Working with a Pharmacy*, p. 7).

Paying for Formula or Supplements

If you need help to pay for formula or supplements, talk with your CF social worker. In some states, Medicaid will cover the cost of formula for people who qualify. The Special Supplemental Nutrition Program for Women, Infants, and Children (WIC) is a government program that, if you qualify, may also be of help to you.

*See CF Words to Know Glossary.



Some insurance companies pay for vitamin and mineral supplements if they are available only by prescription. Some companies that make enzymes offer special programs that will provide some vitamins and high-calorie supplements. Your CF dietitian can tell you more about how your child might qualify.

Making the Transition to Solid Foods

Before you introduce your baby to solid foods, talk with the CF center dietitian or CF doctor. Many babies start on solid foods about the time they are six months old. Plan ahead what you will do about enzymes when your baby is ready for solid food. Talk with your CF health care team about what to do when your child is four to six months old.

Making the Transition to Whole Milk

If your baby is a year old and gaining weight well, he or she is probably ready to switch from formula or breast milk to whole cow's milk. You will need to talk with the CF center dietitian or CF doctor so he or she can help you make any needed change in your child's enzyme dose. The dietitian or doctor can tell you how much milk your child should drink every day. Plan ahead with your team when to change your child to whole milk and how to make the change.

Feeding Your Child

Every parent has trouble at times feeding a baby or toddler. Young children often have times when they are not interested in eating or only want to eat certain foods. You may have special problems feeding your child with CF. You will want to use problem solving to handle these problems. Problem solving is a skill you learn as you Watch and Discover and Think and Act.

If you are worried that your child is not eating enough, talk with your CF health care team. Don't push or force food on your child. Eating and mealtime behavior could easily become a battle between frustrated parents and their toddler. The CF health care team can help you figure out if your child needs more calories and how he or she might get them most easily. You may find it helpful to work with a child psychologist if you continue to struggle with your child's eating behavior.

You can learn more about helping your child cooperate in the module *Working with Your Child*. Another resource you might find helpful is the book *Child of Mine: Feeding with Love and Good Sense* by Ellyn Satter (3rd Edition, 2000).

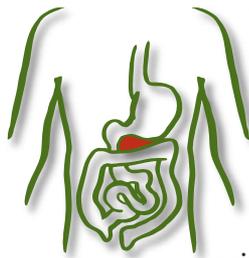


MANAGING YOUR CHILD'S MALABSORPTION

Most people with CF have trouble digesting and absorbing the calories and nutrients from the food they eat. This problem is called malabsorption. To grow and gain weight, people with CF need to take enzymes when they eat. You will learn how to give your child enzymes and how to WATCH AND DISCOVER symptoms of malabsorption.

CF AND DIGESTION

The pancreas (a gland behind the stomach) makes enzymes that digest food in the small



intestine. The enzymes break down fat and protein. Cystic fibrosis often causes the ducts (tubes) going from the pancreas to the small intestine to be blocked with abnormal mucus. If the tubes are blocked, the enzymes cannot reach the small intestine. Without the help of the enzymes, formula, breast milk, and any foods that contain protein and/or fat are not digested well. As a result, the body cannot absorb the fat, protein, and calories from the **intestines*** and is unable to use these foods for energy and growth. (For more information, see “CF FACTS — THE DIGESTIVE SYSTEM” in Appendix 3.) When children with CF are unable to digest their food very well, they have malabsorption. A child with malabsorption may have poor nutrition, and problems growing and fighting infections.

How severe malabsorption is varies from one person with CF to the next, and can change over time. Some people with CF do not have malabsorption. Sometimes the CF health care team will order lab tests to check for malabsorption. Usually, though, the team will depend on you (and your child, as he or she gets older) to watch for symptoms of malabsorption.

The CF health care team will work with you as you learn how to:

- Watch and Discover the signs and symptoms of uncontrolled malabsorption.
- Think and Act to make a plan for what to do when you notice signs and symptoms.

You can learn more about managing malabsorption in the module *Managing Nutrition and Digestive Problems*.



WATCH AND DISCOVER

MALABSORPTION

Signs and Symptoms of Malabsorption

Even though your child will be under the care of the CF health care team, you will need to Watch and Discover to identify signs and symptoms of malabsorption. Sometimes you or the CF health care team may need to adjust your child's enzymes. Symptoms you can watch for include:

- Change in the number of stools
- Change in the appearance of stools
- Poor weight gain or weight loss
- Excess gas
- Bloating
- Cramps and abdominal pain
- Increase in appetite
- **Rectal prolapse***

■ Change in the Number of Stools

Watch to see how often your child has **bowel movements*** (stools) each day. Undigested food makes larger and more frequent stools than normal. You will notice you have to

*See CF Words to Know Glossary.



change diapers more or that your child may have to make many trips to the bathroom.

Most infants and toddlers have one to three stools per day. Breast-fed babies may have a stool with nearly every feeding. More stools than usual for your child is a sign of malabsorption.

■ Change in the Appearance of Stools

The look or appearance of the stool can also change with malabsorption. Changes to watch for include:

- **Size** — Larger stools.
- **Loose stools** — Stools that look bulky and soft and are not well-formed may be due to malabsorption. The stool is not usually watery with malabsorption (see **DIARRHEA***, p. 19). Many children do not have well-formed stools until they are two years old. Look for a change in what is usual for your child.
- **Greasy or floating stools** — The stool may look as though it has grease or oil in it. If your child is toilet trained, the stool may also float. These are signs that your child's body is not able to absorb enough fat.
- **Odor** — Stools may smell worse than usual.

If you notice any of these signs or symptoms, talk to the CF health care team and they may need to adjust your child's enzymes.

Other Signs and Symptoms of Malabsorption

■ Poor Weight Gain or Weight Loss

If the body does not absorb enough calories, a child will lose weight or will not gain weight as expected. You should look at your child's growth chart during visits to your CF center and primary care doctor to make sure your child is gaining enough weight.

■ Excess Gas

This may be the first change you notice with malabsorption. Bacteria that normally live in our **large intestine*** (or **colon***) break down undigested food from the small intestine. This makes gas (**flatus***). The more food that is left undigested, the more gas that is made. The gas may also smell bad.

■ Bloating

With malabsorption, the child's **abdomen*** may become distended, which makes the belly look more round and bloated. Excess gas and extra stool in the intestine causes the bloating. Bloating can make your child feel full. When this happens, your child may not eat enough. Sometimes your child's belly will also feel firmer than usual.

Note: Bloating caused by gas is different than the temporary bloating caused by having a full tummy. After infants or children eat, their bellies look round and full. This fullness goes away as they digest the milk or food. When gas causes bloating, the bloating does not start while the child is eating and it does not go away in the hours after the meal.

■ Cramps and Abdominal Pain

Your infant may seem uncomfortable or more fussy. You may notice this more during or after your baby eats. Young children may not be able to tell their parents when they have a stomach-ache. Sometimes you can tell when your child is uncomfortable and his or her belly hurts, especially before he or she has a bowel movement.

■ Increase in Appetite

Children's appetites normally change a little from one meal to the next and from day to day. Look for longer-term changes. If your child seems to be eating a lot more than before it may be a symptom that he or she has malabsorption and is losing calories. Even though your child may be eating more, his or her hunger is not satisfied because the food is not digested well. If you notice a change in your child's appetite, look for the other signs and symptoms listed here.

■ Rectal Prolapse

In rectal prolapse, part of the **rectum*** protrudes out of the **anus***. The rectum is the very end of the large intestine that is connected to the anus (the hole where stool comes out). When a child strains to push stool out, the rectum may push out. Poor muscle tone and large, bulky stools that are hard to pass cause prolapse. Children





younger than three have rectal prolapse more often because of poorly controlled malabsorption. Although it can be frightening to see, usually it is not dangerous. The rectum often goes back in by itself when the child relaxes or it can be gently pushed back in. If it does not go back in, you need to have your child checked right away. If your child has rectal prolapse, talk with your CF doctor to find out how to prevent it.

Sometimes the symptoms of malabsorption are similar to acute diarrhea or **lactose intolerance*** (inability to digest a sugar found in milk and other dairy products). Here are some clues that can help you and the CF health care team figure out if another problem is causing a change in digestive symptoms.

DIARRHEA

Sometimes parents are confused about whether their child is getting too few enzymes or having acute diarrhea. Diarrhea is not treated by adjusting enzymes.

Diarrhea is most often caused by infection with certain bacteria or **viruses*** and is a common illness among young children. In both diarrhea and malabsorption, you often see a greater number of stools in a day. Diarrhea usually creates different changes in a child's stools than those you will see with malabsorption. In diarrhea, the stools are watery and runny, rather than large and bulky.

Here are some clues that your child has diarrhea:

- Many watery stools
- Friends or family members have the same illness
- Symptoms appear suddenly rather than gradually
- Symptoms last one to two days
- Fever (usually more than 100.4°F)
- Stomach pain
- Nausea (stomach upset)
- Vomiting (this does not happen with malabsorption unless there is a severe **blockage*** in the intestines)

If you notice any of these clues, it is more likely that your child has infectious diarrhea. If your child does not have any of these signs or symptoms, it is more likely that your child has a problem with malabsorption.

Antibiotics can sometimes also cause diarrhea. In this case, there will be many watery stools, but it is unlikely your child will have a fever. The diarrhea will usually stop when the antibiotics are stopped. If you think your child is having diarrhea because he or she is taking an antibiotic, talk to your CF center nurse or doctor. If the symptoms are not too serious, the CF doctor may want you to continue the antibiotics. The doctor may have a suggestion about how to prevent the diarrhea from getting worse. If the diarrhea is severe, the CF doctor may want to change your child to another antibiotic.

Diarrhea can lead to **dehydration*** (not enough water in the body). Children younger than two who have diarrhea are more likely to get dehydrated than are older children. **Dehydration is a serious problem.** Your CF doctor may need to see your child, depending on your child's age and the amount of diarrhea. When your child has diarrhea, do not give him or her just water or clear liquids for a long time because your child could lose weight. If you are not sure what to do, check with your CF center nurse or doctor.

LACTOSE INTOLERANCE

Milk and other dairy products have protein and fat, which provide good calories and nutrients for children with CF. (Many nutritional supplements also contain milk or milk solids.)

Some people have problems digesting the sugar or lactose in cow's milk. Cystic fibrosis does not cause lactose intolerance. In fact, it is a common problem among all children. Children with and without CF can have lactose intolerance.

Tell your CF doctor if you notice your child has diarrhea, gas, and/or bloating right after eating or drinking milk and other dairy products that

*See CF Words to Know Glossary.



WHAT YOU CAN DO

Knowing My Child's Usual GI Symptoms

Use the worksheet "My Child's Usual **Gastrointestinal*** (GI) Symptoms" in the back pocket of this module to help figure out your child's **baseline symptoms*** (including your child's usual pattern of stools) with the dietitian or other CF health care team member. Write down what is usual for your child so that you, and others who help care for your child, can WATCH AND DISCOVER to see if there is a change.

If you think your child has new or increased symptoms, you should talk with the CF health care team. Sometimes it takes days to decide if there is a real change, but if you see a pattern that is different, it is always better to check with the team. Other helpful information you can report includes:

- Change in diet: The team will want to know if there have been any recent changes in your child's diet.
- Enzyme therapy: The CF health care team will also want to know if you are having any problems giving your child enzymes or if you have had a change in enzyme brand or dose.

Using a Symptom Diary

You may find it helpful to keep a diary or chart of your child's stool pattern and any digestive symptoms. This can help you look at the pattern over time and see if there is a problem and if it is related to any specific type of food, activity, or time of day.

You can record:

- Foods your child ate
- Number of enzymes
- Symptoms
- What you did



Making notes on what you observe can be very helpful. When you have made a change in enzyme dose, write it down and watch to see if it has helped your child. You may also want to keep a diary for a week before your next CF visit. Although some people keep a record every day, this is not necessary for everyone. You can decide how a diary may help you work with your child and the team and when you want to keep it.

contain cow's milk. It may be hard to tell if your child needs to take more enzymes, or if your child is having trouble digesting lactose. Symptoms of lactose intolerance can be similar to those of malabsorption. Your CF doctor can do a test to check for lactose intolerance. The CF dietitian and doctor can tell you how to manage lactose intolerance.



THINK AND ACT

TREATING MALABSORPTION

Pancreatic Enzymes

Many people with CF need to take pancreatic enzymes because the enzymes the pancreas makes cannot get to the small intestine. Enzymes come in several brands and formulations. Enzyme products are made as capsules full of powder or beads and your child will take them by mouth with breast milk, formula, or food. The CF health care team will work with you to find the best type of enzymes to meet your child's needs.

The enzymes your child takes travel into the stomach and then pass into the small intestine. Once in the small intestine, the enzymes help digest the food your child has eaten. Nutrients from the digested food can then be absorbed into the body.

Giving Enzymes

Give enzymes just before your child eats or drinks. Talk about enzymes with your child as you give them. Even toddlers can begin to see that enzymes are a normal part of every meal and snack. You might say, for example, "Time for your enzymes." or "Where are your enzymes?" or "Let's get your food helpers." Encourage your child to ask for enzymes when he or she is eating and then praise your child for asking.

Enzyme Powder: Sometimes you may need to use powdered enzymes. Enzyme powder should be mixed with a small amount of breast milk or formula before giving it to your baby.



Babies may not be able to swallow enzymes unless they are sucking at the same time. Make sure your child drinks something right after taking the enzymes. Enzyme powder can irritate the gums and tongue if it remains in the mouth. If you breast-feed, be sure to wipe out your baby's mouth beforehand to remove any extra enzymes. Enzymes can irritate your breast.

Enzyme Beads: Young children and infants who cannot swallow capsules can take the enzyme beads. Open the capsule and mix the beads inside with 1 to 2 teaspoons of soft foods that your child does not have to chew (for example, applesauce or baby food fruits). Try not to use a food with fat or protein that will activate the enzymes before your child swallows them. Ask your CF dietitian which foods work the best to mix with enzyme beads.

Your child should swallow the beads right after they have been mixed with food. Always give the enzymes before your child eats a meal or snack. The beads should not be chewed or crushed. They will not work as well if they are chewed. Enzymes can irritate your child's mouth and affect his or her **teeth enamel**.* Make sure your child drinks something right after taking the enzymes. Again, if you breast-feed, be sure to wipe out your baby's mouth to remove any extra enzymes so they do not irritate your breast.

Enzyme Capsules: Children aged four and older (and sometimes as young as three) can learn to swallow enzyme capsules. You can find more information about working with your child to take enzymes and dealing with problems you may have with cooperation in the module *Working with Your Child*.



Avoiding Generic Enzymes

Generic* enzymes are not designed for treating CF. Generic enzymes are not as reliable and may not control malabsorption as well as brand-name enzymes. A pharmacy may try to substitute a generic enzyme for the enzyme

your doctor has prescribed. Even if the cost may be lower, the generic enzymes are not a good choice. Do not change the brand of enzymes without talking with your CF health care team.

Storing and Refilling Enzymes

Check your supply of enzymes to make sure you know when you need to order a refill. Do not keep enzymes in extreme hot or cold areas. Check the expiration date on the bottle to make sure the enzymes are fresh.

How Many Enzymes to Take

Unlike most medicines, how many enzymes your child needs does not depend on how old your child is or how much he or she weighs. Instead, it depends on how much the ducts between the pancreas and the small intestine are blocked. As children grow and get older, how much the ducts are blocked can change. The more blockage your child has, the more **replacement enzymes*** your child will need to take.

Younger children with CF usually take the same amount of enzymes with every meal. They also take a set number of enzymes with every snack that has fat and/or protein. You will work with the CF health care team to find out the amount of enzymes your child needs.

Older children with CF sometimes need to adjust the number of enzyme capsules they take with meals and snacks. When they eat meals that are larger or higher in fat than usual, they take more than the "usual" number of enzymes. For example, a pizza with cheese and meat has more fat and protein than a lower-fat food like turkey. Changing the number of enzymes to match what your child eats may seem complicated at first. You will learn more in the module *Managing Nutrition and Digestive Problems* and by working with your CF health care team.

Your child will not need to take enzymes with some foods, especially foods that only have **carbohydrates*** (for example, fruit, fruit juice,

*See CF Words to Know Glossary.



Jell-O). Your CF dietitian can help you learn what foods your child can eat without taking enzymes.

Enzymes and Diaper Rash

Some of the pancreatic enzymes can pass through in your child's stools and cause a diaper rash. The rash tends to be around your child's anus. Change your baby's diaper right away. When you first notice your child's bottom is irritated, cover the area with petroleum jelly or zinc oxide at every diaper change. If diaper rash continues to be a problem, talk to your CF doctor. The doctor can prescribe a special cream to help protect your child's bottom. Your doctor may also need to adjust your child's enzymes. Your child can have other kinds of diaper rash just like any other baby. Check with your primary care physician for other types of diaper rash and how to treat them.

Teaching Other Caregivers How to Give Enzymes and Watch for Malabsorption

Your child may have other people who help care for him or her during mealtimes (a grandparent, day-care center, or babysitter). They will need to understand how to watch for malabsorption and how to give enzymes. Be sure other caregivers know that your child will need enzymes before meals and snacks.

WHAT YOU CAN DO

You can put your child's enzyme doses on the worksheet "What to Feed My Baby" or "What to Feed My Child" in the back pocket of this module. You may have different doses for meals and snacks. Ask your CF dietitian or doctor if you can make any adjustment based on what your child eats or what you WATCH AND DISCOVER. Share this worksheet with everyone who helps feed your child.



You can also make a chart and check off the doses of enzymes with your child each day to help show what a good job he or she is doing. This chart might also include other "jobs" the child does each day, such as brushing teeth or helping get dressed.

Day-care centers and schools will treat enzymes as any other medicine. You will need to sign forms, bring a pharmacy-labeled bottle, and leave written instructions. You may also need to bring applesauce or other food to mix the enzymes with.

Work out a system for other caregivers to tell you about your child's dirty diapers and stools. Many day-care center staff have daily report forms, which include a record of bowel movements. Be sure to tell the staff to also write down the size and appearance of the stools and show you the records.



MAKING IT WORK FOR YOU

TIPS ABOUT ENZYMES

Remembering Enzymes

For some families, remembering enzymes can be a problem at times. Here are some suggestions from other families:

- Take time to get organized. Think of ways to remind yourself.
- Put the enzyme bottle near where your child eats and where you will notice it.
- Write yourself a note and post it on the refrigerator.
- Keep enzymes in your purse, backpack, or in your child's diaper bag. Always have some with you.

After you have used these cues or signals for a while, giving enzymes will become a habit. In time, you and your child will connect mealtimes and snacks with giving enzymes.

These ideas are examples of strategies that parents have come up with when they have used problem solving. Problem solving is a skill you will learn as a CF manager. Problem solving is a part of Think and Act. You will learn more about problem solving in the module *Becoming a CF Manager*.



MANAGING YOUR CHILD'S LUNG PROBLEMS

Cystic fibrosis makes **lower respiratory tract infections*** more likely and harder to get over. The lungs can have problems from abnormal mucus and **inflammation***. Some lung problems can be prevented. By watching for signs and symptoms of infection, families can find and treat infections sooner. Managing CF includes preventing and treating lung problems to help delay lung damage.

There are three major causes of lung problems in CF:

1. **Obstruction*** — **airways*** become blocked with abnormal mucus
2. Inflammation — airways become irritated and swollen
3. Infection

All three of these work together in a cycle to cause lung damage over time. To manage and prevent lung problems in CF, you and the CF health care team will take steps to manage and prevent airway obstruction, inflammation, and infection. In this section, you will learn:

- How to watch for signs and symptoms of infection
- Ways to help prevent infection and manage the abnormal mucus obstructing or blocking the airways

You will be able to learn these skills and more by working with your child's CF health care team and from the modules *Becoming a CF Manager* and *Managing Lung and Other Respiratory Problems*.

LUNG PROBLEMS

The respiratory system is divided into the **upper respiratory tract*** (**sinuses***, nose, mouth, and throat), and the **lower respiratory tract*** (**windpipe*** or **trachea***, airways, and lungs). Most of the lung problems in CF start in the airways of the lower respiratory tract. The airways are the tubes that carry air to and from the lungs. The airways also help fight infection and keep **irritants***, such as dust or smoke, from getting deeper into the lungs.

To learn more about the airways and parts of the respiratory system, see “CF FACTS — THE RESPIRATORY SYSTEM” in Appendix 4.

Bronchitis* and **bronchiolitis*** are names for infections in the lower airways (**bronchi*** and **bronchioles***). **Pneumonia*** is an infection of the **lung tissue***. Infections that start in the airways can spread in the lung and cause damage. Over time, repeated infections can reduce how well the lungs function and can shorten a person's life.

CYCLE OF LUNG PROBLEMS

- **OBSTRUCTION:** The airways get obstructed or blocked with abnormal mucus. The abnormal mucus is a good place for bacteria to live and cause infection.
- **INFLAMMATION:** The airways get swollen and irritated—and this causes more mucus to be made. Chemicals released from inflamed cells can cause damage to the lungs.
- **INFECTION LEADS TO MORE INFLAMMATION AND MUCUS:** When there is an infection, there is also more inflammation in the lungs as the body tries to fight the infection. The airways then produce more mucus as well.
- **REPEATED INFECTIONS LEAD TO DAMAGED AIRWAYS:** Damaged airways make it even harder to clear out mucus. Mucus builds up and the bacteria grow causing more infection. Infection causes damage, and lung problems get worse.



*See CF Words to Know Glossary.



WHY IT IS IMPORTANT TO PREVENT RESPIRATORY INFECTIONS

Not all **respiratory infections*** can be prevented, but some can. Fewer infections means there will be less damage to your child's lungs. Your doctor will help you keep the number of infections your child has to a minimum.

Treating an infection *early* can also help prevent permanent lung damage. To help treat infections as soon as possible, the CF health care team will work with you to:

- Watch and Discover symptoms of infection
- Watch and Discover changes using lung health tests at clinic visits
- Think and Act with the CF health care team to make a CF action plan to prevent and control respiratory problems

Solve problems using skills learned from the CF FEP program and your CF health care team when following your child's respiratory treatment plan or CF action plan.



WATCH AND DISCOVER

LUNG PROBLEMS

Signs and Symptoms

You can look for signs and symptoms that your child may have an infection. If you know what to look for, you may be able to find the problem early and get treatment started.

The early signs of infection may be hard to detect. In the beginning, an infection, such as bronchitis, may cause only a small change in symptoms. Unless you are watching for these changes, you might not notice them until the infection is more serious. With practice, you can begin to recognize signs and symptoms of a lower respiratory tract infection early. Watch your child for these signs and symptoms:

- Cough
- Increased mucus
- Tiredness
- Decreased appetite
- Weight loss or failure to gain weight

- Irritability
- Fever
- Changes in breathing—such as your child feeling short of breath, breathing harder, or breathing faster

You will want to talk with the CF health care team about what the best clues are for your child. Here is a closer look at each of these signs and symptoms.

COUGH

Some people think children with CF always have a cough. This is not true. Some children have a chronic cough and others do not. Watch for when your child starts to cough, or if there is a change in your child's usual cough. Notice:

- Sound (For example, Is it deep? Wet?)
- Frequency (How often? How long?)
- When (Morning, night, with activity, with treatments?)
- Severity (How much does it bother your child?)

What this may mean: A wet or loose cough means there is more mucus. A dry, harsh cough may mean the airway muscles are tight and are causing narrowing (this is called **bronchospasm***). A cough that starts when your child lies down may be caused by postnasal drip or congestion in the upper airways (sinuses and nose).

What to do: If you notice a change in cough, look for other symptoms. Check your CF action plan to see if you need to start or increase any treatments. Check in with your CF nurse to see if you need to bring your child to the clinic.

INCREASED MUCUS

Babies and toddlers usually cannot spit out the mucus or **sputum*** they cough up from their lungs. Instead, they swallow it. This means you cannot look at the mucus as you would with an older child. Sometimes a young child will cough so hard that he or she will vomit up mucus and then you can see how thick the mucus or sputum looks.



What this may mean: With infection, a child's mucus usually gets thicker and stickier, and may change color. Rather than clear or white, as it is normally, mucus can become yellow or green.

What to do: Look for other signs of infection. Check your CF action plan to see if you need to start or increase any treatments. Check in with your CF nurse to see if you need to bring your child to the clinic.

TIREDFNESS

Look for changes in your child's energy level and activities. Notice:

- Tiredness
- Sleeping more
- Restless (cannot get comfortable); your child may be sleeping more but for fewer hours at a stretch
- Playing less, especially if your child is not interested in favorite activities

What this may mean: Tiredness may mean that the body is using more energy to fight an infection.

What to do: Look for other signs of infection. Check your CF action plan to see if you need to start or increase any treatments. Check in with your CF nurse to see if you need to bring your child to the clinic.

DECREASED APPETITE

Any child's appetite may vary from meal to meal and from one day to the next. Young children may also go through times when they eat only one or two favorite foods. *Notice if your child is eating less over a longer period of time or has little interest in food (even favorite foods).*

What this may mean: Your child's appetite can decrease when your child has an infection. Sometimes if your child has swallowed quite a bit of mucus, his or her stomach can become upset and he or she may not feel like eating.

What to do: Watch closely for other symptoms. If you notice other signs of infection, talk to the CF health care team and start your CF action plan.

WEIGHT LOSS OR POOR WEIGHT GAIN

Your CF health care team and primary care doctor will watch your child's weight gain at clinic visits. At home you can notice if your child has:



- Lost weight
- Not gained weight for some time
- Looks thinner
- Has not changed clothing sizes for a long time

What this may mean: Children with infections often lose their appetites. Even if your child is eating the same amount, it takes calories to fight infection. Your child will have fewer calories to help keep up his or her weight. Another reason may be that your child is having increased malabsorption.

What to do: Look for other symptoms of infection. If you notice other signs of infection, talk to the CF health care team and start your CF action plan. Also, watch for other symptoms of malabsorption (see *Watch and Discover for Malabsorption*, pp. 17–19).

IRRITABILITY

From time to time, you may notice that your child is "just not acting like herself." Notice if your child is:

- Fussy
- Frustrated easily
- Withdrawn
- Aggressive

What this may mean: Changes in behavior or attitude may mean that your child is not feeling well physically. Some changes in behavior may be part of your child's growing older, or they could be caused by stresses in your child's environment. You may want to take a closer look at what is going on in your child's life. (Is your child teething? Has your child started going to a new day-care center or has your family schedule changed?) Behavior change, however, can be an early clue that a child is getting sick.



What to do: Watch your child closely for any other symptoms of infection. If you notice other signs of infection, talk to the CF health care team and start your CF action plan.

FEVER



When your child feels warm to the touch or looks flushed, take his or her temperature. (For help, see “TAKING YOUR CHILD’S TEMPERATURE” in Appendix 6.)

What this may mean: A fever usually means there is an infection somewhere in the body. An infection in the lungs can cause fever, but children with CF do not always have a fever when they have a lung infection or lower respiratory tract infection. Your child could have bronchitis or pneumonia even if he or she does not have a fever.

What to do: Look for other signs of infection. If there are other signs along with the fever, contact your child’s CF health care team and start your CF action plan. If there are no signs of a lower respiratory tract infection, follow the directions from your child’s primary care physician to look for other kinds of infection (such as a cold or ear infection) and when you need to bring your child in to be checked. Follow the doctor’s instructions on how to treat your child’s fever.

CHANGES IN BREATHING

Changes in breathing include feeling short of breath, working hard to breathe, and breathing faster than usual. Watch for these changes in how your child breathes:

- The skin around your child’s ribs and/or neck pulls in with each breath. (This is called **retractions*** and is a sign your child is having to work hard to breathe.)
- Breathing faster than usual at rest or while sleeping

To decide if your child is breathing faster than usual, you will need to know what your child’s usual breathing rate is at rest and asleep.

You can use the worksheet “My Child’s Usual Respiratory Rate” in the back pocket of this module to figure out your child’s resting **respiratory rate***.

What this may mean: When your child has a lower respiratory tract infection, his or her lungs have to work harder. If the airways are blocked, it is harder for the lungs to move air in and out. The lungs may not be taking in as much oxygen. This can make your child breathe faster. Your child will have to use more muscles in the chest and rib cage to breathe, which causes retractions. Your child may look short of breath to you. Young children usually cannot tell you how their breathing feels. Problems with breathing are more often seen when an infection has gotten worse or more severe.

What to do: If you think your child is having difficulty breathing, call your CF center right away. If your child is turning blue or having a great deal of trouble breathing, call 9-1-1 or take your child to the nearest emergency room. Tell the emergency room team that your child has CF and have them talk with the CF doctor on call.

If you are not sure that your child is having trouble breathing, count your child’s breathing rate for 1 minute when he or she is resting quietly or asleep. If it is higher than usual, check it again in 15 minutes, and then in one hour. If the rates are high, call the CF center and start your CF action plan.

ALLERGY OR INFECTION?

At first, it may be hard for you to figure out whether, for example, your child’s cough means bronchitis, or a cold, or an allergy. The symptoms of an allergic reaction (runny nose, coughing) can resemble the symptoms of a cold. People with allergies are sensitive to certain things in the environment, such as pollen, mold, or animal dander. Allergy problems tend to run in families. Other family members who have allergies could be a clue



that your child may also have allergies. Sometimes, though, parents think allergies are causing their child to cough because they don't want to admit to themselves that their child has bronchitis. But remember, if your child does have bronchitis it needs to be treated right away to prevent lung damage. You may want to have your child tested for allergies, so that you have more information when you need to make decisions about symptoms. Talk to your primary care physician or CF doctor to see if your child needs allergy testing.

On the other hand, if your child does have allergies, an allergic reaction can make CF problems worse. During an allergic reaction a person's upper and/or lower airways become inflamed or swollen. Allergies can cause increased mucus that can lead to more problems in CF. Treating allergic reactions is important in controlling CF respiratory problems. You can learn more about this in the module *Managing Lung and Other Respiratory Problems* and by talking with your CF health care team.

COLD OR LOWER RESPIRATORY TRACT INFECTION?

You may find it hard to decide when your child has just a cold (an **upper respiratory tract infection***) or when he or she has a lower respiratory tract infection. Sometimes what starts as a cold can move into the lungs.

With a cold there is usually little or no fever. A high fever may suggest a more serious infection. In CF, a child can have a lung infection without a fever. A cough can be due to either an upper or lower respiratory tract infection.

If your child has a new cough (especially a wet, deep-sounding cough); is coughing more often; or has rapid breathing, retractions, shortness of breath, or **wheezing***, he or she could have a lower respiratory tract infection.



If you are not sure whether your child has more than a cold, bring your child to your primary care physician's office or CF center. If your child is not better after three to five days of treatment, check back with your primary care physician or CF health care team.

LUNG HEALTH TESTS

Along with the symptoms you and the CF health care team will watch for, there are tests that the CF health care team will sometimes do to look for a lung problem. These include:

- **Sputum** (mucus) **cultures***
- Chest radiographs (**X-rays***)
- **Oxygen saturation*** levels
- **Lung function tests***

Some of these tests, such as the lung function tests, your doctor will do at regular checkups. Sputum cultures are done one or more times every year to see if any bacteria are growing in the mucus in the lungs. Chest X-rays are done at least once a year and as needed when your CF doctor suspects any changes. Your CF health care team will help you decide what tests your child needs. Each of these tests adds information, much like putting together the pieces of a puzzle. The test results, along with your child's symptoms, will give you and the team a more complete picture of how your child's lungs are doing.

Sputum Culture

Babies and young children with CF often do not cough or spit out mucus. To get a sputum culture, the CF health care team does a "gag" culture. A throat swab is used to tickle the back of the throat, which can make the child gag. As the child gags, sometimes mucus will come up from the lower windpipe and the swab can be used to catch it in the back of the throat. Although this sounds uncomfortable, it can be done quickly and does not hurt. The CF health care team does this test to find out what kind of bacteria may be causing the problem. Different bacteria are treated with different antibiotics.

*See CF Words to Know Glossary.



Chest X-Rays

Even if the doctor does not hear any new sounds in your child's chest with a stethoscope, the doctor may be able to see changes in the airways on a chest X-ray. A chest X-ray can show chronic and acute changes, such as air being trapped by blocked airways or pneumonia.

Oxygen Saturation Levels

The oxygen saturation level tells the CF health care team how well your child's lungs are taking in oxygen. Sometimes with a lung problem, the oxygen saturation level will be lower than normal, even if a child does not look like he or she is having problems breathing.

Lung Function Tests

Lung function tests show the CF health care team how well your child's lungs are moving air in and out. With infants, the CF health care team will need to use **sedation*** (a medicine to help your child go to sleep) and special equipment to do the lung function test. As a child gets older, the CF health care team will teach him or her how to blow into a computer that measures lung function.

You can learn more about these tests in the module *Managing Lung and Other Respiratory Problems*.

WHAT YOU CAN DO

Your Child's Usual Respiratory Symptoms

Once your child is stable and symptoms are under control, you can figure out your child's usual or baseline respiratory symptoms. If your child is in the hospital, you will want to wait until he or she is better.

To decide if your child is showing early signs of infection, you need to know what is "normal" for your child. What is your child like on a usual day when he or she is well? For many children this will mean they have no symptoms at all. For other children, a usual day may mean they cough every morning.



Knowing your child's everyday symptoms will help you notice any changes. Use the worksheet "My Child's Usual Respiratory Symptoms" in the back pocket of this module to record your child's usual or baseline pattern of symptoms. Watch your child for a week and look for the symptoms listed on the chart. Talk with other people who regularly spend time with your child to see what they notice. You may be surprised by how differently each of you describes the same symptoms. You may have different ideas about what is normal for your child or for a child of the same age. You may not agree on what "a lot" and what "a little" is. Talk with each other and the CF health care team to reach an agreement on how to describe your child's CF symptoms.

Take the chart with you to your next visit to the CF center. Go over these everyday symptoms with your child's CF doctor.

Make copies of your child's baseline symptom chart and give them to other caregivers. Ask them to let you know about any differences they see in your child—both good and bad.

Your Child's Usual Respiratory Rate

Your child's respiratory rate is simply how fast your child breathes. All children have different respiratory rates when they are resting or asleep and when they are active. The respiratory rate is not always the same even at rest—usually you will have a normal range rather than a single number. Older children breathe more slowly than babies or young children. For example, a 1-month-old infant often breathes 30 to 40 times per minute at rest, whereas a 1-year-old child usually breathes 15 to 20 times at rest. A young child's change in breathing rate is more obvious with a respiratory infection than it is in an older child. Use the worksheet "My Child's Usual Respiratory Rate" in the back pocket of this module to figure out your child's normal range.

Using a Symptom Diary

Some parents find it helpful to keep a record of their child's symptoms every day. You can use a symptom log or diary to write down what you notice. To keep a log or diary, you can write down your child's symptoms and the date on a piece of paper or in a notebook. Using a symptom diary helps you keep track and makes it easier to



tell the CF health care team what you have noticed. You may find it helpful to even write down days without symptoms if you are trying to decide how a medicine is working.

Some people keep a record all the time. Others write down symptoms from time to time. You can use a symptom diary to help you figure out your child's usual symptoms when he or she is feeling well. You can also use a diary to help you remember what you want to report for clinic visits. A diary may be a very helpful way for you to track how your child is responding to a change in treatment or a new medicine.

You can use "My Child's Usual Respiratory Symptoms" worksheet, or you can use the comment section of your baseline symptom form.



THINK AND ACT

PREVENTING INFECTION

Although it is not possible to avoid all infections, there are ways you can try to protect your child from some infections. Infections are caused by germs, including viruses and bacteria. Certain bacteria can cause infection in people with CF more than in other people. Children with CF can also have more trouble with the same infections that other children get.

The most important ways to prevent infection are:

1. Good hand washing
2. Immunizations
3. CF checkups
4. Good nutrition
5. Physical activity

Note: Physical activity and good nutrition can actually help your child resist or fight infection. Active play is a child's exercise and exercise can help strengthen the lungs and move mucus out of the airways.

Here are the steps you can take every day to limit the number of respiratory infections your child gets.

Hand Washing

Most respiratory infections are spread by person-to-person contact. Surprisingly, most viral infections are not spread through the air. Instead, they are spread through direct contact with people's hands or by touching surfaces **contaminated*** with germs.

When a person coughs, sneezes, or rubs his nose, germs wind up on his hand. If that same person then touches a toy, a nipple on a bottle, a child's hand, or anything else that winds up in the child's mouth or nose, the bacteria or viruses have hitched a ride. Germs can also take a ride directly from your hand to your child's mouth or nose without stopping off anywhere in between.

To stop germs from spreading, wash your hands before you give your baby a bottle or before you feed your child. Wash your child's hands, too. Ask other caregivers and visitors in your home to wash their hands before they handle your child. You will find that alcohol-based hand sanitizers work just as well as soap and water. Carry a small bottle of alcohol-based hand sanitizer with you in your purse or diaper bag and use it often.



In day-care centers, it is very easy for adult hands to give germs a ride from one child to the next. Most centers have policies about hand washing for their staff. Make sure the day-care workers are aware that your child is at higher risk of infection. Get your child's classroom a large container of alcohol-based hand sanitizer and ask that the teacher have other children use it, too.

*See CF Words to Know Glossary.



Avoiding Ill People

You want your child to play with friends, yet you want to be careful. Your child should avoid close contact with adults and children who have colds or other respiratory illnesses until their symptoms are gone. Ask parents to let you know if their child is ill before making a play date.

If you know you have a viral infection, be especially careful to wash your hands after you sneeze, cough, or rub or wipe your nose. You can wear a mask, but that does not completely prevent the virus from spreading.

The CF center has policies about hand washing during clinic visits and for cleaning clinic rooms between patients. The CF center also has isolation rules to restrict contact with people in the clinic or hospital who have certain kinds of infection. The CF health care team can tell you more about these rules and how they apply to your child and protect against the spread of infection.

Avoiding Air Pollution

Indoor and outdoor air pollution are common irritants that many people are exposed to. Irritants are things in the air that are harmful to the lungs when they are breathed in. Irritants can cause inflammation and injure airways.

Tobacco smoke is a form of air pollution that you can avoid. Protect your child from exposure to cigarette, cigar, and pipe smoke, as well as other types of smoke and vapor fumes from **electronic nicotine delivery systems (ENDS)**,* such as e-cigs. Ask everyone, including grandparents, day-care providers, and other caregivers, not to smoke in your home, or around your child. If you or anyone else in your home smokes, now is the time to stop. There is no safe exposure for anyone. For



more help, see “KEEPING YOUR CHILD HEALTHY - ELIMINATING TOBACCO AND NICOTINE SMOKE EXPOSURE” in Appendix 7.

You have less control over the outdoor air. Air pollution can sometimes be a risk to people who have lung problems from chronic diseases, such as **asthma*** or CF. One common air pollutant is ozone. If ozone is a problem in your area, there is a warning system that can help you decide when to have your child avoid outdoor play. You can check reports on air quality for your area in the local news. You can also get air pollution and ozone air quality information about your area on the United States government AIRNOW Web site at <http://airnow.gov>.

Getting Immunizations

Many “childhood illnesses” are serious respiratory infections. There are immunizations that will protect your child from some of these infections, such as **Haemophilus influenzae*** type b (Hib) and **Streptococcus pneumoniae*** bacteria that can cause pneumonia and other infections. Ask your child’s primary care doctor for a schedule of when your child should have immunizations. Because your child has CF, it is even more important for your child to have this protection.

One illness that is important to protect your child against is **influenza*** (the flu). The influenza virus can quickly cause pneumonia once a person starts to get sick with the flu.

Your child should get a flu **vaccine*** every year. A child as young as six months can start taking the flu shot. If your baby is too young to get the flu vaccine (less than six months old at the time flu vaccines are given), it is important that those who care for your baby get immunized. The nasal flu vaccine is *not* recommended for people with lung problems.



THINK AND ACT

Children and adults who are between the ages of 2 and 49 years old, and who do NOT have lung problems, can take the nasal flu vaccine. It is a good idea to have everyone in your household get a flu vaccine each year. Check with your primary care physician in October about getting a flu vaccine.

The flu vaccine prevents most flu illnesses, but if your child has flu symptoms even after having had the vaccine, call your CF health care team. Flu symptoms often include a fever of 102°F or higher early in the illness. There are anti-viral medicines that can treat the flu infection early and make it less likely to cause serious illness. If you think your child has the flu, you should call the CF health care team right away.

Your CF health care team will let you know if there are any other special immunizations your child should have to prevent infection.

Antibodies to Prevent RSV Infection

Another important virus to try to protect children with CF from is called **RSV*** (respiratory syncytial virus). This virus often causes cold symptoms, but can cause pneumonia and bronchiolitis. Babies who have CF and are less than two years old may receive palivizumab shots (brand name Synagis®) to protect against RSV infection. The shots are not quite like other immunizations. Most immunizations help your body make its own **antibodies*** to fight a specific infection. This is not yet possible for RSV. These shots give the baby antibodies to help fight RSV infection. The shots are given monthly during RSV season. Talk with your CF or primary care doctor about whether your child needs these shots and when they would be given. Before the RSV season, your doctor can also help you get prior authorization from your insurance company to find out how much it will cover the cost of the shots.

MANAGING AIRWAY OBSTRUCTION

Obstruction, or blocked airways, is a common problem in CF. Obstruction causes problems with air flow and makes it harder for a person to breathe. It can also make the lungs more prone to infection. Obstruction happens mostly in the small airways and can be hard to hear even with a stethoscope. Abnormal mucus causes airway obstruction in CF. The airways can also narrow when the airway muscles go into spasm and become inflamed and swollen. To manage and prevent airway obstruction in CF, you need to learn how to keep mucus out of your child's airways by using airway clearance and breathing treatments.

Airway Clearance

Airway clearance is a term that refers to techniques you can use to help loosen mucus in your child's airways so that your child can cough it up and out of the lungs. A common airway clearance technique is called **chest physical therapy*** or **CPT*** (referred to in the past as **postural drainage and percussion*** or **PD & P***). CPT uses **percussion*** and **vibration*** to loosen and move mucus. It is most often used with infants, but it can be done with a person of any age.

Percussion, also called "clapping," uses a cupped hand or a small plastic device to briskly pat the chest and back. (With a small baby, the motion can resemble burping, but the motion is more forceful than that.) Clapping loosens the mucus and helps move it out of the lungs.

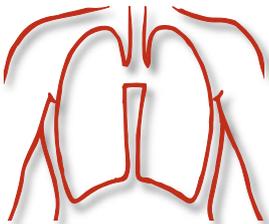
Vibration is another action that is sometimes used with CPT to help loosen mucus. Vibration is a quivering movement of the hands on the chest to help gently shake the mucus into the larger airways. Vibration needs to be timed

*See CF Words to Know Glossary.



with the child breathing out, so it normally is not used with a small child. Vibration may be suggested for some older children. Your CF health care team can tell you if it would be helpful to do some vibration along with CPT.

As you care for your child, you eventually will learn about other ways of clearing the airways that do not require as much help from you or other caregivers. One example is a **percussive vest*** that helps loosen mucus by shaking the chest after the child puts it on. Some methods require more cooperation from the child, such as those that use **autogenic drainage*** and **active cycle of breathing*** techniques, or the **Flutter® device*** or **acapella® Vibratory PEP Therapy System.***



With any of the techniques to clear out the airways, you will want to encourage your child to cough or **huff*** up the loosened mucus. A young child will not spit mucus out. But, even if he or she swallows it, the mucus is still out of the lungs. So if you don't actually see any mucus or sputum coming out of your baby's mouth, it doesn't mean the therapy is not working.

Do airway clearance before feeding your child or wait at least one hour after he or she has eaten. If your child has a problem with throwing up during or after airway clearance, talk with your CF health care team. If you are doing chest physical therapy, you may need to change some of the positions. Airway clearance is usually done after breathing treatments that help loosen mucus. You should ask the CF health care team in what order you should do breathing treatments and airway clearance.

A therapist, or other member of the CF health care team, will show you how to do airway clearance. Often children need to do airway

clearance every day to help clear mucus from their lungs. The team will help you sort out:

- How many times a day to do airway clearance (how many sessions your child needs)
- When to do airway clearance
- What techniques and positions to use and in what order to do them
- How long to do airway clearance during a session
- How much to increase airway clearance when your child is ill

Even if you and your child do not normally do airway clearance, your doctor will ask you to start when your child has a lower respiratory tract infection—especially if your child's cough is more frequent, his or her chest sounds congested, or his or her cough sounds wet with more mucus. If you do CPT, your doctor may suggest focusing more on one position or one area of the lung. If your child starts to have more frequent lower respiratory tract infections, your doctor may ask you to start or to increase the number of airway clearance sessions you do with your child. Ask for a review at CF clinic visits if you are not sure you are doing the airway clearance correctly.

See "AIRWAY CLEARANCE: DOING CHEST PHYSICAL THERAPY WITH YOUR INFANT OR TODDLER" on pp. 37-44. This section includes illustrations of common CPT positions. You can review the positions as you learn to do airway clearance and photocopy them for easy reference when you teach others who care for your child.

Respiratory Treatments

Respiratory treatments are also called breathing treatments. Liquid medicines are turned into a mist by a **nebulizer*** with a medical **air compressor.*** The child breathes the mist into his or her lungs by wearing a **face mask*,** which covers his or her nose and mouth. If the child is older, he or she uses a





mouthpiece* Other medicines are released as a mist from **metered dose inhalers*** (called MDIs) and are used with a valved **holding chamber*** (**spacer***) with a face mask.

Different medicines may be prescribed for breathing treatments. Some medicines help open the airways, some thin mucus, and others help reduce swelling and inflammation in the airways. All of these are ways to relieve airway obstruction. Your child's CF doctor will decide which medicines you should use with your child's respiratory treatments.

Some children do respiratory treatments every day. Others do them only when they have an infection. Your child's CF doctor will work with you to decide what your child needs.

If your child needs respiratory treatments, a member of the team will show you how to do them.

During treatments, it is important that you hold the mask on your child's face. If the mask is away from your child's face, you will lose much of the medicine in the air and it will not work as well or last as long. Your child should be sitting up for the treatment to get the most benefit. For more information on giving respiratory treatments, see "HOW TO DO BREATHING TREATMENTS WITH YOUR INFANT OR TODDLER" in Appendix 8.



MAKING IT WORK FOR YOU

CHILD COOPERATION

By age two or three your child can help in small ways, such as:

- Coughing during or after treatments. (Coughing is important even if nothing is coughed up.)
- Learning to turn quickly to the next position.
- Taking deep breaths and **exhaling*** while you do vibration.

Everyone has trouble sometimes with a child cooperating with treatment. In young children this can be a normal part of growing up. Children between the ages of 18 and 24 months often do not want to cooperate. To learn more about helping your child adjust to breathing treatments and airway clearance, see the module *Working with Your Child*.

If your child continues to cry with most treatments, ask the CF health care team to help you figure out why and what you can do.



THINK AND ACT

TREATING INFECTION IN CF

You have Watched and Discovered changes in symptoms that may mean your child has a respiratory infection. You will want to Think about what is causing the symptoms and what treatment your child may need. You will work with your CF health care team to Act to treat the infection and make sure the symptoms get better.

Lower respiratory tract infections, such as bronchitis, are usually treated with:

- Respiratory treatments
- Airway clearance
- Antibiotics

You will use one or more of these treatments when your child has an infection. You may also use one or more of them every day to help prevent your child from getting infections.

Respiratory treatments and airway clearance help fight infection by opening up blocked airways and moving mucus out of the lungs. This is important because bacteria grow in airways blocked with mucus and can cause infections. Infection causes more mucus to be produced. Mucus that blocks the airways can make it harder to breathe.

*See CF Words to Know Glossary.



Antibiotics are medicines that kill or control bacteria. The doctor will prescribe antibiotics to treat lower respiratory tract infections caused by bacteria.

Antibiotics

Children will take one or more antibiotics when they have an infection. Some children will also take antibiotics every day or every other month to prevent further infections.

To decide which antibiotic(s) to use, your doctor will consider:

- The types of bacteria that usually cause specific infections in children with CF and which antibiotics work the best in fighting those bacteria.
- The antibiotics that have done the best job of fighting infections in your child when he or she was sick before.
- Any **side effects*** or allergies your child has had from specific antibiotics.
- The types of bacteria growing in a sample of your child's mucus (sputum). In the laboratory, the sputum is cultured (put on special gels that bacteria can grow in) to find out which bacteria are in the sputum. The lab can even test different antibiotics to see which ones do the best job fighting the bacteria in the culture.
- How the antibiotics will be given to your child. Antibiotics may be given by mouth (**orally***), by **aerosol*** mist breathing treatments (inhaled), or through the vein (**intravenously*, IV***) depending on the type of antibiotic and your child's needs. In some cases, the bacteria are more sensitive to antibiotics that are given by IV. For some infections, using several antibiotics together works better than one alone.

Giving Your Child Antibiotics

When your child is on oral antibiotics, follow these guidelines. You may also find it helpful to review "GIVING YOUR CHILD MEDICINE" in Appendix 9.

- **Give the right amount at the right times.** There must be enough (or right level) of the antibiotic in the blood and lungs to fight the infection. But too much medicine can cause problems as well. Be sure to give the right amount of medicine at the right times.

Measure correctly. Make sure you give the right dose of medicine each time.

Keep on schedule. Different medicines work for different periods of time. If you are late by just an hour, give the dose. If you are late by more than an hour, wait until the next dose time. *Ask about any other special instructions for giving the medicine.* For example, your child must take some medicine with food, but other medicines your child must take on an empty stomach. Some medicines your child should not take at the same time. You can keep track of your child's antibiotic doses on the worksheet "My Child's Antibiotic Dose Tracker" in the back pocket of this module.

- **Give all the medicine.** Antibiotics must be taken for the total number of days prescribed—even if the signs of infection are gone. Antibiotics may make your child feel better quickly. When this happens, it may be hard to remember to give the rest of the medicine. But if your child does not take all the medicine, the bacteria may grow again or become resistant to the antibiotic. The second infection may be harder to control than the first one.
- **Get all lab tests done that your doctor orders.** For some antibiotics, your doctor may order lab tests to check the **drug level*** in your child's blood. The test results will help make sure that your child is getting the right amount of the drug. Other tests may be done to check for side effects, such as kidney irritation.
- **Watch for side effects and allergic reactions.** The most common side effects of antibiotics are diarrhea and stomach upset. The most common sign of an allergic reaction to an antibiotic is a rash. If the diarrhea is mild, you may decide to continue with the antibiotic and deal with the side effect. If the side effects are severe, or if you think your child is having an allergic reaction to the drug, call your CF doctor's office. **Do not stop an antibiotic without talking with your child's CF doctor first.** ("My Child's Antibiotic Dose Tracker" worksheet also has a column for you to note side effects.)



You may find it hard at times to get your child to take an oral antibiotic. The antibiotic may not taste good or your child may not like the texture. Sometimes toddlers may simply



resist cooperating with taking medicine (especially if they can tell that it is upsetting to their parents).

You can do several things to help solve these problems. But your child is most likely to cooperate with taking antibiotics if, from the very beginning, you are consistent and matter-of-fact about it.

- **Give the antibiotic with a small amount of food your child likes.** Offer something good to drink after the medicine. Check with your CF health care team or pharmacist to make sure you

can give the antibiotic with food. Ask the pharmacist if a flavoring can be added to the medicine—the pharmacy has syrups that can be used to flavor liquid medicines.

- **Praise your child for cooperating with taking medicine.** Positive reinforcement usually works much better than punishments or threats.

If you find that taking antibiotics or other medicines is becoming a daily problem, talk with the CF health care team. They may have suggestions. For more information about helping your child take medicines, see the module *Working with Your Child*.

WHAT YOU CAN DO

Your Child's CF Action Plan

Your child's CF health care team will give you a written CF action plan. This plan tells you what to do every day to manage your child's lung problems in CF. Think of it as your battle plan for preventing and fighting infection. Also, it will have instructions for how you will increase treatments when you notice a change in symptoms. For example, you may increase the number of airway clearance or breathing treatments. You can find samples of CF action plans in the back pocket of this module.

The treatment plan you work out with your child's CF doctor will usually include:

- What treatments to do every day on a routine basis
- When to add other treatments
- What other treatments you should do
- How often to do them
- When and how much improvement to expect
- When to contact the CF health care team
- When to return to your regular schedule and treatments

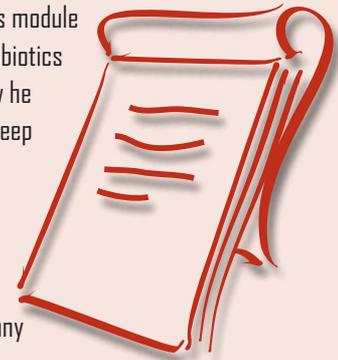
You should watch to see how your child's CF action plan is working. If you start a treatment plan for infection, your child should show some improvement within three to five days. If your child does not improve, or if your child's symptoms get worse, call your CF health care team.

When the treatment is scheduled to stop, your child should be back to his or her normal or baseline symptoms. If your child still has unusual symptoms, continue the CF action plan until you talk with your CF health care team. You can find more about using CF action plans in the module *Becoming a CF Manager*.

Your Child's Antibiotic Record

Use the worksheet "My Child's Antibiotic Record" in the back pocket of this module to write down which antibiotics your child takes and how he or she reacts to them. Keep a written record of your child's:

- Types of antibiotics
- Side effects
- Allergic reactions, if any



Having a written record will help as you work with the CF health care team. For example, if you have to talk to a doctor on call at night who does not know your child well, you will want to remember which medicine your child usually does not tolerate.

Put your medicine record and CF action plan in a CF travel folder. You can learn more about using a CF travel folder in the module *Becoming a CF Manager*.



SUMMARY

Beginning CF Care gives you an overview of basic CF management and how to work with your child's CF health care team. In this module you have learned the basics of monitoring for respiratory and digestive symptoms, using common treatments to control and prevent malabsorption, managing airway obstruction, and preventing or treating respiratory infections. These are skills that you and your family will need to practice and do with your child. As you gain more experience, you will become more confident and eventually become the expert in your child's care. This is exactly what the CF health care team hopes will happen. You are an important partner with the CF health care team. As your child grows and matures, the CF health care team will help you teach him or her to take more responsibility and become a partner, too.

You will want to continue to work with the CF Family Education Program to learn more. The other modules in this program will have more details about becoming a CF manager and how you can work out nutrition, digestive, and respiratory problems. Stories from other families of children with CF (called "Learning from Other Families") can help you find out what has worked for them in different situations. One of the modules will help you work with your child as he or she grows. Review this material from time to time and as your child's needs change. Share it with others who also care for your child. Ask questions, and don't be afraid to let others know when you need more information. Learning to live with a chronic disease is the best way to keep it in good control.

The CF health care team, along with valuable information and tips from these modules, will help you become a successful CF manager and an expert in your child's care.





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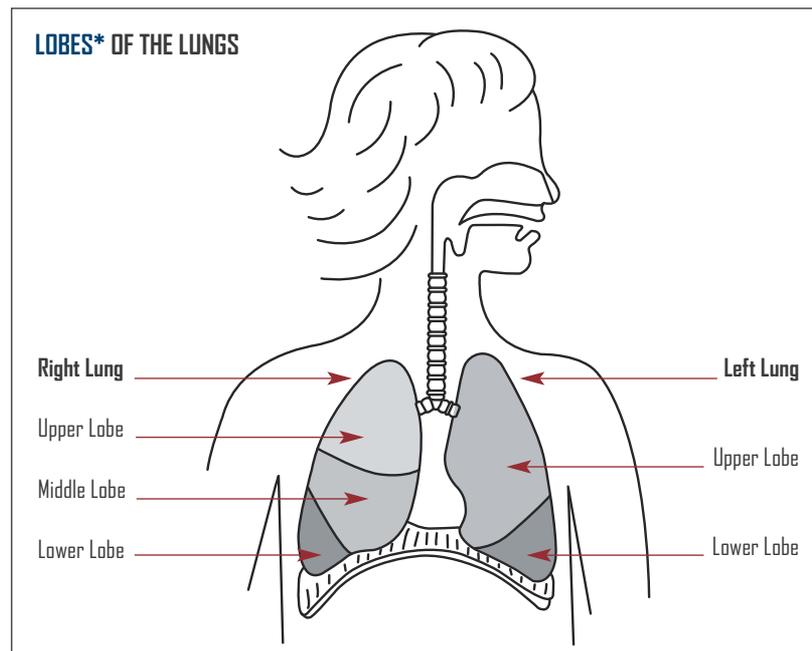
AIRWAY CLEARANCE: DOING CHEST PHYSICAL THERAPY WITH YOUR INFANT OR TODDLER

Chest physical therapy or CPT (referred to in the past as postural drainage and percussion or PD & P) is a common type of airway clearance used in infants and toddlers with CF. Chest physical therapy helps loosen mucus in the airways so the child can cough it up. Even if your child changes to other airway clearance methods in the future, such as a **percussive vest***, knowing how to do CPT is important as a backup.

The main goal of all types of airway clearance is to have the child cough mucus out of the airways. Babies and young children do not cough out mucus often, but even if they cough it up to the throat and swallow it, the mucus is still out of their lungs. Swallowed mucus can pass through the intestines into the stool.

NOTE TO PARENTS:

- Do airway clearance before feeding your child or wait one hour after he or she has eaten. Talk with your CF health care team if your child has problems with throwing up. If you are doing CPT with your child, you may need to change some of the positions.
- Airway clearance is usually done after breathing treatments that loosen mucus. Ask your CF health care team in what order you should do breathing treatments and airway clearance.





Correct Hand Position

You may do percussion with your hand or you may be given a plastic clapper or small face mask to use. When doing percussion by hand, your hand should look like this:



- Your hand should be cupped, fingers together.
- Your hand should make a hollow sound as it claps the chest.

If your hand is cupped correctly, percussion should not be painful. To make percussion more comfortable, make sure your child has a thin layer of clothing or cover the area you are clapping with a sheet.

Correct Areas to Clap

Always limit percussion or clapping to your child's rib cage. Clapping on the backbone, breastbone, or below the rib cage will make your child uncomfortable. Always feel the area that you want to clap to be sure that you are in the right position before starting.

TIP: Making Yourself and Your Child Comfortable

So that you and your child are both comfortable during CPT, you may want to hold him or her on a pillow in your lap. You can position your child either across your legs or lengthwise from your waist to your knees (see illustrations of different positions, pp. 40–44). You can also do CPT sitting on a bed next to your child. You want to be comfortable and relaxed so that you do not strain yourself. Your CF health care team can help you find positions that work for you as well as for your child. Some young children even go to sleep while being clapped. You can use some of these same techniques as your child grows older, although the positions will vary a little for the older child.

How Often to Do CPT

The CF health care team will work with you to decide how often to do CPT with your child. You may not be doing all the positions shown in this section. There are blanks under each illustration for you to write in how long and how many times a day you will do each position. Ask your therapist or other CF health care team member to help you fill in this information.

When your child has a cold or respiratory infection, you will want to increase the number of times you do CPT. Again, your CF health care team will help you decide how many times per day you should do CPT when your child is sick.

Some parents find it helpful to do their child's airway clearance positions in the same order each time. This can help you:

- Learn the positions.
- Remember to do all the positions and not accidentally skip one.
- Encourage your child to learn the positions and how to change into the next position quickly.

You can photocopy the positions you and your child use and put them in the order you want to do them.

For more information about different types of airway clearance, see the CF FEP module *Managing Lung and Other Respiratory Problems* or go to the CF Foundation Web site (<http://www.cff.org>), or call 1-800-FIGHT-CF.

*See CF Words to Know Glossary.



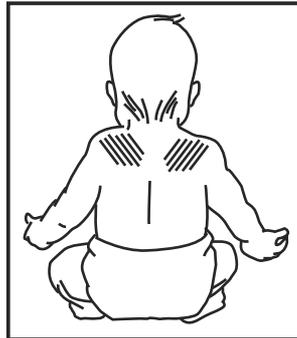
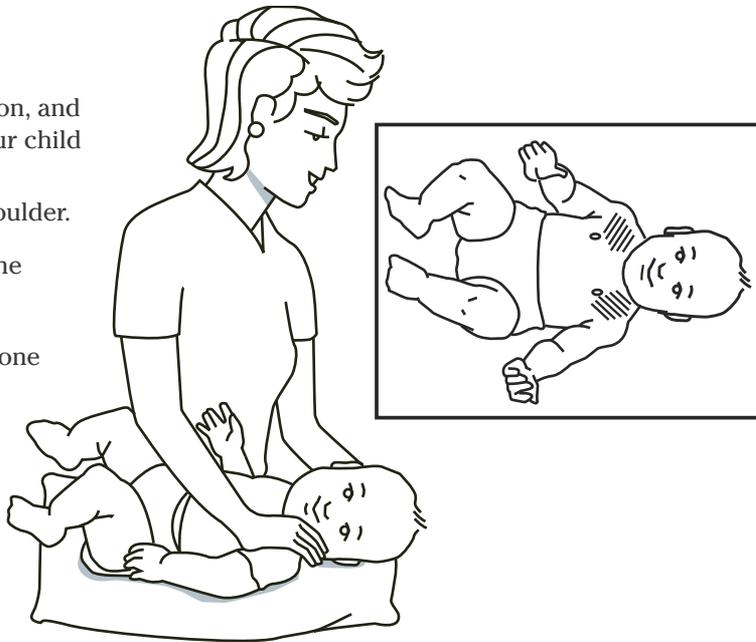
UPPER LOBES, FRONT SEGMENT

Lay your child flat on his or her back.

Note: You can use a pillow for this position, and for others, to help make yourself and your child more comfortable.

Clap between your child's nipple and shoulder.

- Clap under your child's left collarbone for ____ minutes ____ times a day.
- Clap under your child's right collarbone for ____ minutes ____ times a day.



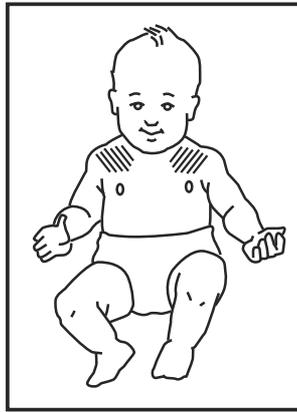
UPPER LOBES, BACK SEGMENT

Your child should sit in your lap, bending slightly forward. Support your child's head and neck with your arm.

Clap over your child's shoulder blade.

Do **not** clap on your child's backbone.

- Clap your child's left shoulder blade for ____ minutes ____ times a day.
- Clap your child's right shoulder blade for ____ minutes ____ times a day.

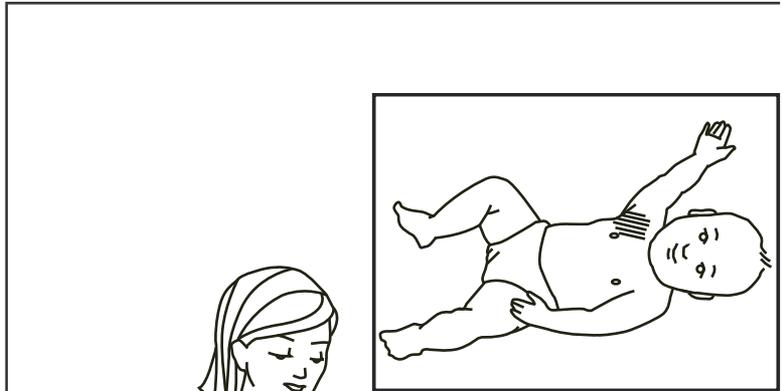


UPPER LOBES, TOP SEGMENT

Cradle your child in your arm and lap. Your child should be about halfway to sitting upright.

With your child's head resting on your arm, clap between your child's nipple and the top of his or her shoulder.

- Clap between your child's left nipple and shoulder for ____ minutes ____ times a day.
- Clap between your child's right nipple and shoulder for ____ minutes ____ times a day.



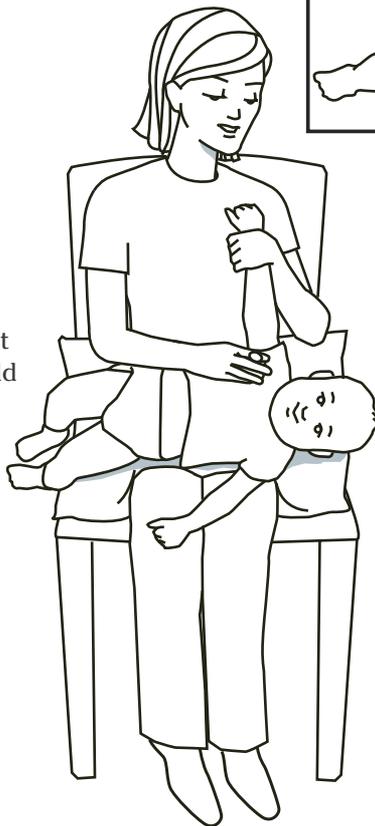
RIGHT MIDDLE LOBE

Put your child flat on your lap on his or her left side. Roll him or her back part way (about a one-quarter turn), but do **not** turn your child onto his or her back.

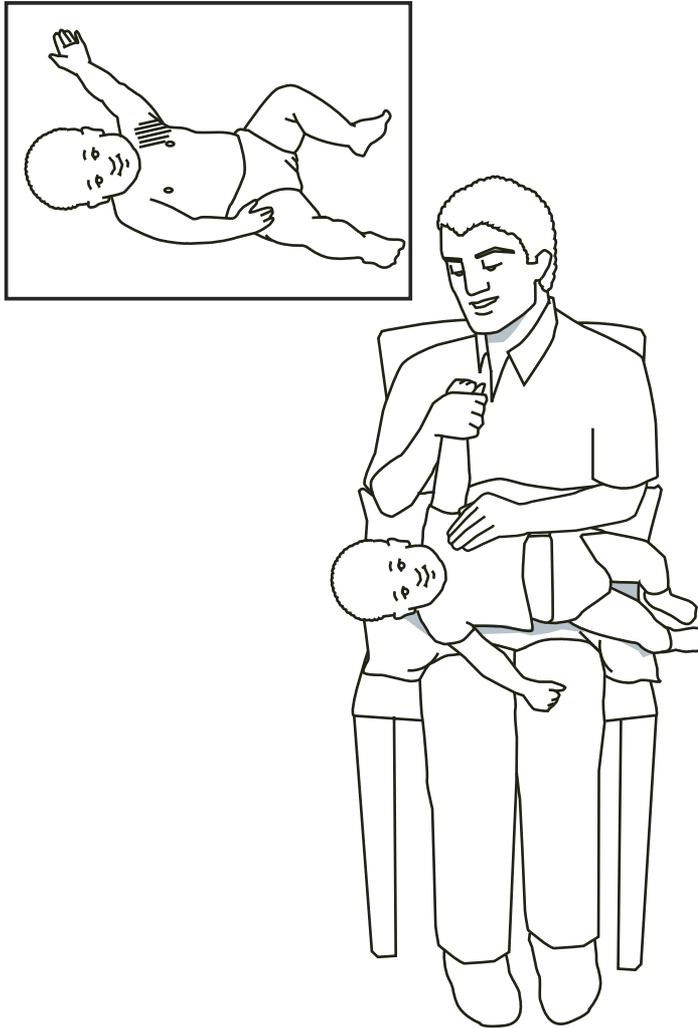
Clap between your child's right nipple and right armpit.

- Clap between your child's right nipple and right armpit for ____ minutes ____ times a day.

*Note: The parent in this illustration is using a **manual percussor**.**



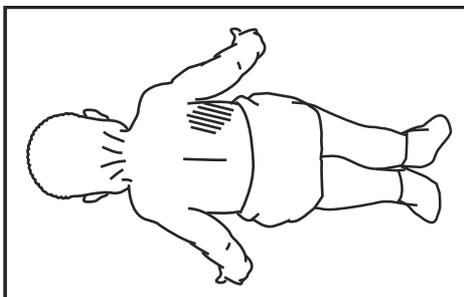
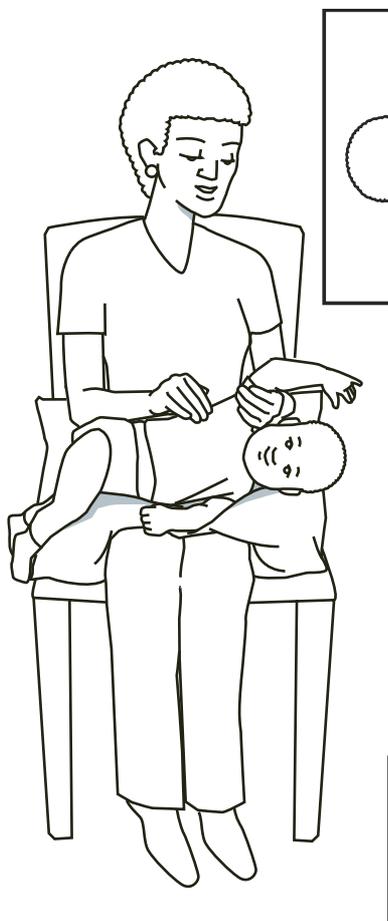
*See CF Words to Know Glossary.



LEFT LUNG SIDE

Put your child flat on your lap on his or her right side. Roll him or her back part way (about a one-quarter turn), but do **not** turn your child onto his or her back. Clap between your child's left nipple and left armpit.

- Clap between your child's left nipple and left armpit for ____ minutes ____ times a day.

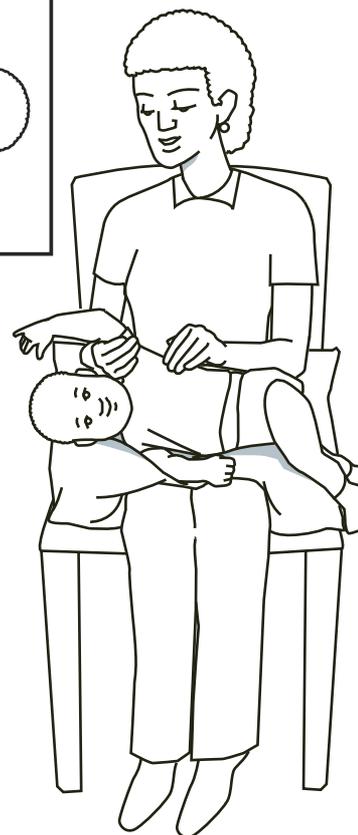
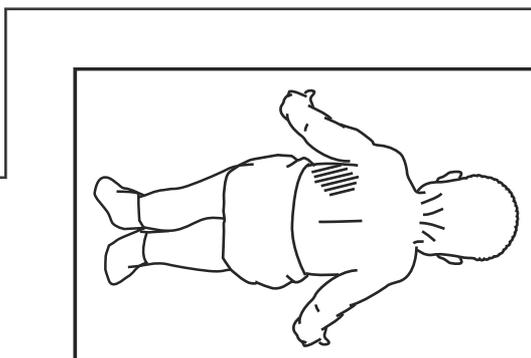


LOWER LOBES, (RIGHT) SIDE SEGMENT

Put your child flat on your lap, with your child lying on his or her left side. Hold your child's arm up above his or her head.

Clap over the side of the chest, just above the bottom edge of the ribs.

- Clap the right side of your child's rib cage for ____ minutes ____ times a day.

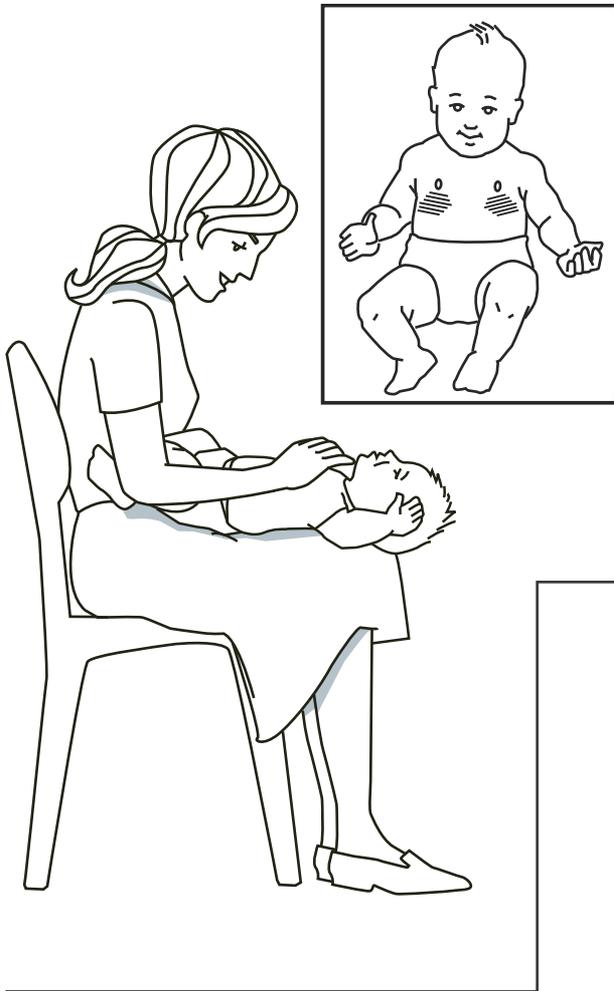


LOWER LOBES, (LEFT) SIDE SEGMENT

Put your child flat on your lap, with your child lying on his or her right side. Hold your child's arm up above his or her head.

Clap over the side of the chest, just above the bottom edge of the ribs.

- Clap the left side of your child's rib cage for ____ minutes ____ times a day.



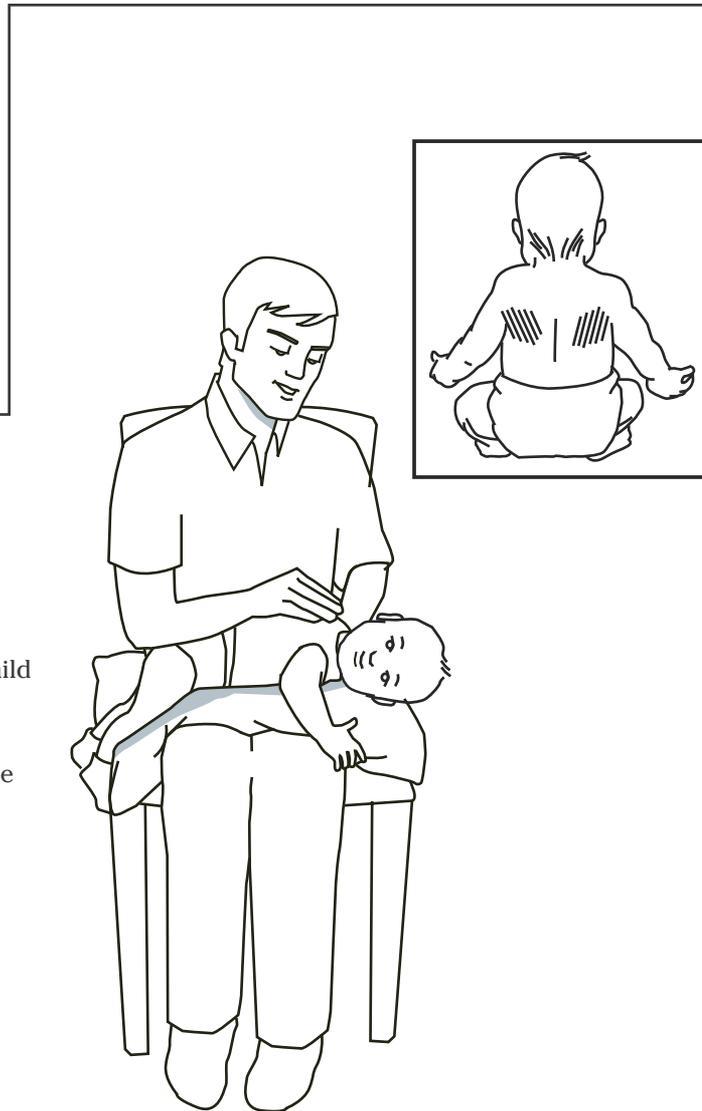
LOWER LOBES, FRONT SEGMENT

Place your child face up flat on your lap.

Clap between your child's nipple and the bottom of the ribs. Do **not** clap on the breastbone.

Do **not** clap on your child's stomach.

- Clap the left side of your child's chest for ____ minutes ____ times a day.
- Clap the right side of your child's chest for ____ minutes ____ times a day.



LOWER LOBES, BACK SEGMENT

Place your child flat on your lap, with your child lying on his or her tummy.

Clap over the side of the back, just above the bottom edge of the ribs. Do **not** clap below the ribs. Do **not** clap on your child's backbone.

- Clap the left side of your child's back for ____ minutes ____ times a day.
- Clap the right side of your child's back for ____ minutes ____ times a day.



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WORKSHEETS

(Located in back pocket)

MY CHILD'S CF HEALTH CARE TEAM

WHO TO CALL, WHO TO VISIT

MY CHILD'S FOOD DIARY

WHAT TO FEED MY BABY

WHAT TO FEED MY CHILD

MY CHILD'S USUAL GASTROINTESTINAL (GI) SYMPTOMS

MY CHILD'S USUAL RESPIRATORY RATE

MY CHILD'S USUAL RESPIRATORY SYMPTOMS

CF ACTION PLAN SAMPLES:

MY CHILD'S CF ACTION PLAN — MANAGING DIGESTION AND NUTRITION

MY CHILD'S CF ACTION PLAN — MANAGING LUNG PROBLEMS

MY CHILD'S CF ACTION PLAN

MY CHILD'S ANTIBIOTIC DOSE TRACKER

MY CHILD'S ANTIBIOTIC RECORD

WHAT TO DO IF MY CHILD HAS A FEVER

"DEAR SMOKER" LETTER



APPENDIX I: THE GENETICS OF CYSTIC FIBROSIS

Cystic fibrosis* (CF*) is a **genetic*** or **inherited*** disease that occurs when both parents pass a CF **gene*** on to their child. Cystic Fibrosis can be found in all races and ethnic groups. Cystic Fibrosis is, however, most often seen in people who are white and who are not of Hispanic ethnicity. At present, about 30,000 children and adults in the United States have CF (about 70,000 worldwide).

Each person has a set of blueprints or instructions found in his or her genes. Genes contain the instructions for how the body develops and works. For example, genes control eye, hair, and skin color. Genes tell the body how tall to grow. Genes determine what blood type a person has. Genes can also cause people to have certain health problems. In all of our body **cells*** we have two sets of **chromosomes***, one from our mother and one from our father. Chromosomes are made up of many genes—they are the holders for genes in the cells. Each cell has 23 pairs of chromosomes that contain thousands of genes. Chromosomes and genes are made of special chemical structures called **DNA*** (deoxyribonucleic acids). The pattern of DNA is what makes up the instruction code in each gene. Genetic testing to look for CF is sometimes called CF **DNA testing***.

Cystic fibrosis is a disease that is caused by an abnormal gene. An abnormal gene is called a **genetic mutation***. The gene that causes problems in CF is found on the seventh chromosome. There are many **mutations*** (abnormal genes) that have been shown to cause CF disease. Over 1000 mutations have been discovered,

but there are about 30 that are common. The most common gene mutation is called **F508del***. When a person receives two CF gene mutations, the person will have CF disease. The cells in the body will follow the instructions from the CF genes and will not work properly.

Which body systems and cells have problems vary with different types of CF gene mutations. For more on what happens to cells in a person with CF, see “WHAT CAUSES PROBLEMS IN CYSTIC FIBROSIS?” in Appendix 2.

HOW CF IS INHERITED

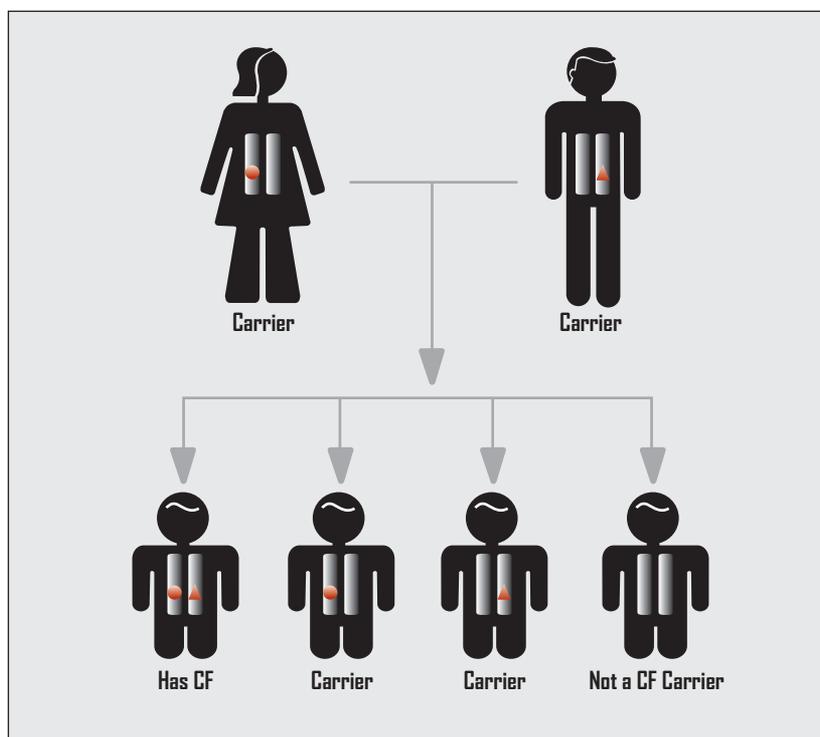
A person must inherit two CF genes to have CF disease. When your child was **conceived***, he or she received a CF gene from both you and your partner. A child can inherit CF only if both parents

carry a CF gene (that is, each parent either has CF or is a **carrier***) and both parents pass the CF gene on to their child. The illustration on this page shows how CF is inherited when both parents are carriers.

There is nothing that parents do to cause CF in their child and usually they do not know that they are carriers of a CF gene.

A carrier is a person who has one abnormal CF gene and one normal gene. Remember, each cell in a person has two of each chromosome, one from the mother and one from the father. If one of the seventh chromosomes has a CF gene and one does not, the person is a carrier and does not have any **symptoms*** or disease problems. This is the way the CF gene is passed on for many generations.

INHERITING CF: BOTH PARENTS CARRYING CF GENE





If your child received one CF gene and one non-CF (normal) gene, he or she would not have CF disease—your child would be a CF carrier instead. The normal gene, not the CF gene, would control what the body’s cells do and would help make sure they work well enough to prevent CF disease. About 1 out of every 25 white Americans is a CF carrier. There are more CF carriers in the white non-Hispanic race than in any other race.

A person can be a CF carrier even though CF disease has not occurred in the family for many generations. This is because a person who is a CF carrier must have a child with someone else who is also a CF carrier and both of them have to pass the abnormal gene to the child. Families may have CF carriers who do not have symptoms and have not had a child who received two abnormal CF genes. Most people who have children with CF did not know that CF genes ran in their families. Cystic fibrosis was only recognized as an inherited disease in the 1930s. Many families may not have known about children who had CF and died in past generations.

WHEN BOTH PARENTS CARRY A CF GENE

When a man and a woman who are both CF carriers conceive a child, one of three things happens:

- There is a one in four chance (25% of the time) the child will receive a CF gene from each parent (two abnormal genes total) and will be born with CF disease.
- There is a two in four chance (50% of the time) the child will receive one CF gene from one parent and a non-CF normal gene from the other parent. This child becomes a CF carrier like the parent.

- There is a one in four chance (25% of the time) the child will receive non-CF normal genes from each parent. When this happens, the child cannot have CF disease and is not a CF carrier.

The illustration on the previous page shows this pattern of inheritance with both parents as carriers. Each time a child is conceived by two CF carriers, the chance that the baby will have CF disease is one in four (25%).

Some parents think that if they have had one child with CF, their other children will be born without the disease. This is not always true. With every pregnancy, parents who both carry the CF gene will always have a one in four (25%) chance of having a child with CF.

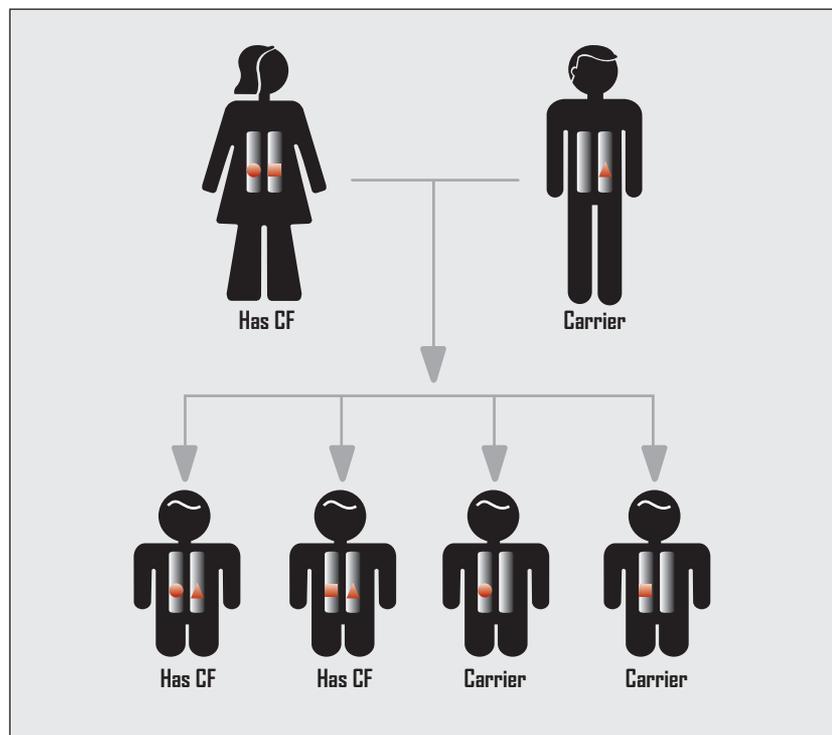
For example, the chance of having a girl is one in two or 50%. You might expect a family of four children to have two boys and two

girls. But you probably know families who have four girls and no boys. Or four boys and no girls. Likewise, two CF carriers could have four children who all have the CF disease.

WHEN A PERSON WITH CF HAS A CHILD

A person who has CF has two abnormal CF genes. A person with CF will always pass an abnormal gene to his or her child. Every child a person with CF conceives will have at least one CF gene. Whether the child is a carrier or has the CF disease depends on the other parent. If a person with CF conceives a child with another person who is a CF carrier, the chance of having a child with CF is one in two (50%). The risk of the child only being a carrier is one in two (50%). The child will either have CF or be a carrier. The illustration on this page shows how CF is inherited when one parent is a carrier, and the other

INHERITING CF: PARENT WITH CF AND PARENT CARRYING CF GENE





parent has CF. If a person with CF conceives a child with a person who is not a CF carrier, the child will always be a CF carrier (100%), but will not have CF.

People with CF have problems with **fertility*** and may not be able to conceive a child naturally. To learn more about fertility and reproduction in CF, see “PUBERTY AND FERTILITY IN CF” in the appendix of the module *Managing Nutrition and Digestive Problems*.

GENETIC TESTING

A person can have a genetic test to see if he or she is a carrier of a CF gene. The genetic test for CF mutations is usually done using a person’s blood sample. Sometimes this is done when a woman is considering getting pregnant or early in her pregnancy. There are ways to test a baby before birth to see if the baby has abnormal CF genes. Some tests only look for the most common mutations and might miss a rare CF gene. Other tests can look at the seventh chromosome for every kind of CF gene. This test is usually more expensive and takes longer to get results. If people already know family members who carry the CF gene, they can make sure the CF test is done to find out if they also carry the gene.

Cystic fibrosis carrier testing may be something to think about in a family who has a child with CF. If there are other children who do not have CF, parents may want to know if they are carriers who could pass the CF gene on to their own children. Cousins, aunts, uncles, and other relatives may also want to know if they are CF carriers. A person with CF who is thinking about having a child may

want to consider having his or her partner tested to know better what the risk will be of having a child with CF.

A child can be **diagnosed*** with CF from CF genetic testing. If a child is found to have two abnormal CF genes, he or she will have the disease. Even if the child does not have symptoms at the time the test is done, it would be expected that the child will have problems from CF in the future. Cystic fibrosis varies from person to person in the types of problems it causes and when symptoms first appear. Some babies are born with symptoms and most will have some **signs*** of CF disease in the first year of life. But other people with CF may not have problems until later.

If the specific gene mutations for a child are known, the family can use that information to help look at the pattern of genes in the family. This can be most helpful if the child has rare mutations that might not be tested for in a usual genetic screening test.

FUTURE RESEARCH

At this time, knowing the gene mutations a person has does not tell us much about how severe a child’s disease may be. Genetics researchers have found some mutations to be milder. People who have these mutations do not have the digestive problems seen with the more common CF mutations. Even so, there are differences among those with common mutations; the types of problems and when they occur still vary from person to person.

Current **research*** on new treatments may benefit some people with CF depending on what CF genes they have. Scientists continue to develop and test

medicines that could help the cells with any CF gene mutations.

Researchers are also looking at other genes that may either help a person who has CF have less severe problems or will make the person’s disease more severe. These genes may modify or alter how CF genes work in the body. Scientists are still learning more about how genes work in our bodies.

Scientists are also working on ways to give a person with CF copies of normal genes in the hope that the normal genes will help the cells work normally. Researchers are testing various gene treatments or **therapies*** that could help organs most affected by CF (such as the lungs). In **gene therapy***, the scientist inserts a normal gene into a cell. But this type of therapy is still very much in the testing phase and it is not clear when it will be available to people with CF.

GETTING MORE INFORMATION

Your CF health care team can tell you more about genetic testing, including carrier and **prenatal*** testing. Many families find it helpful to talk with a **genetic counselor*** about the pattern of inheritance and risks of having another child in the family with CF. This information may help families make a choice about having more children, having genetic testing, or both. Any member of your family, such as a child, niece, nephew, sister, or brother who is considering having a child, can also ask for carrier and prenatal testing and counseling.



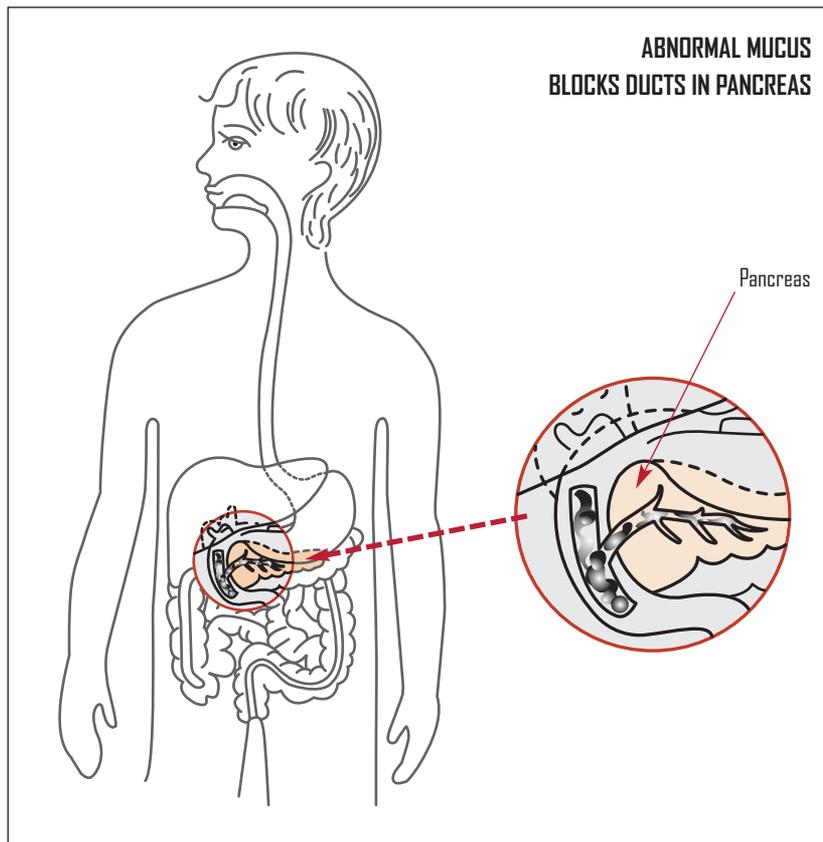
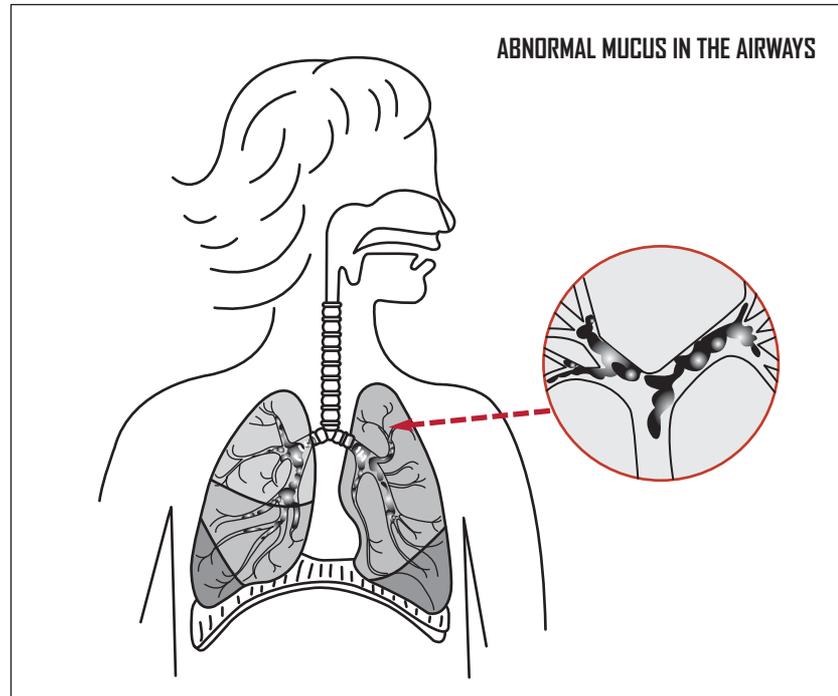
APPENDIX 2: WHAT CAUSES PROBLEMS IN CYSTIC FIBROSIS?

Doctors and researchers have known for a long time about the symptoms of **cystic fibrosis*** (**CF***). Through research, they have begun to understand what causes these symptoms.

ABNORMAL MUCUS

Many of the symptoms in CF are caused by abnormal (thick, sticky) **mucus*** in the body. It is hard for the body to move this mucus.

- Abnormal mucus forms in the **airways*** and is hard to cough up. When mucus stays in the **lungs*** it makes a good place for **bacteria*** to grow. The bacteria cause **lung infections*** that over time can damage the lungs. Some CF treatments help clear mucus from the lungs.



- Mucus blocks the passages (**ducts***) in the **pancreas*** that connect to the **small intestine.*** The pancreas makes **enzymes*** the body uses to **digest*** food in the small intestine. When these passages are blocked, the enzymes cannot get to the small intestine to do their job. Without these enzymes, the body does not digest food as well. **Calories*** from **fat*** and **protein*** in food are not **absorbed*** and are lost in the stool. The person with CF cannot grow and gain weight normally. This is why most people with CF have to take **replacement enzymes*** when they eat.

The abnormal mucus that people with CF have is the result of a faulty set of instructions being used by the cells.

*See CF Words to Know Glossary.



CELLS AND GENES

Cells* are the very small, basic working units of all living things. The human body is made up of billions of cells. They are so small that they cannot be seen with the naked eye. Some cells make up the skin. Other cells form the heart, **liver***, muscles, and bones. Cells are what actually do the work in the body. If the body makes something (a tear, an enzyme, a **hormone***), it is made by certain cells. The cells even do the work to help turn food into energy the body can use.

Each cell needs instructions on what it is supposed to do and how it should do it. These instructions come from the **genes***. Genes are simply coded messages that tell the cells how to do their jobs. Human beings have thousands of genes. Some genes decide what color eyes or hair will be. Other genes decide blood type. Every cell contains many genes. The genes are arranged on **chromosomes***. You can learn more about genes and chromosomes in “THE GENETICS OF CYSTIC FIBROSIS” in Appendix 1.

THE CFTR GENE

One particular gene carries the instructions for making the **CFTR*** protein. CFTR is short for *cystic fibrosis transmembrane conductance regulator*. We all have this gene and we are all supposed to have the CFTR protein in our cell walls. But in people with CF, the gene is abnormal. This is called a **gene mutation***. The instructions are different. There are several ways that the instructions are changed with an abnormal CF gene. Some people with CF have cells that do not make any CFTR protein. In some people, the CFTR protein is not made correctly or it does not end up in the right place in the cell to do its job. These differences cause the problems in CF.

THE CFTR PROTEIN AND CF

The CFTR protein normally functions as a passage (called a channel) in the cell wall. The CFTR channel allows **chloride*** to go through the cell wall. Chloride is a part of salt and is important to the body’s cell chemistry. The movement of chloride in and out of the cell also affects how water and sodium go in and out of the cell. This is important in cells that produce mucus. Salt and water flow plays a role in how much mucus is made and how thick it is.

Scientists have found that even if the CFTR protein is present in the cell walls, it is not working or works poorly depending on the gene mutations. People with CF have this problem in a number of different cells in their bodies, including the cells in the **sweat glands***, **lungs***, pancreas, and intestine. The abnormal CFTR protein explains why people with CF have high chloride levels in their sweat (salty sweat). The high level of chloride in sweat has been known for a long time and is why the **sweat test*** is used to help diagnose CF.

Scientists are doing research on how the CFTR protein works and how to develop ways to improve chloride movement across the cell wall. Cells can have other problems that cause symptoms in CF, but the abnormal CFTR protein is considered the primary problem in CF. Researchers continue to try to understand more about how CF affects the body and the functions of its cells. Ask your CF health care team and learn more about FDA approved medicines that help the CFTR protein work. Scientists continue to develop and test other medicines that could help the cells with other gene mutations. As researchers find out more about what causes CF, their discoveries can help lead to new and better treatments.



APPENDIX 3: CF FACTS – THE DIGESTIVE SYSTEM

THE GI TRACT

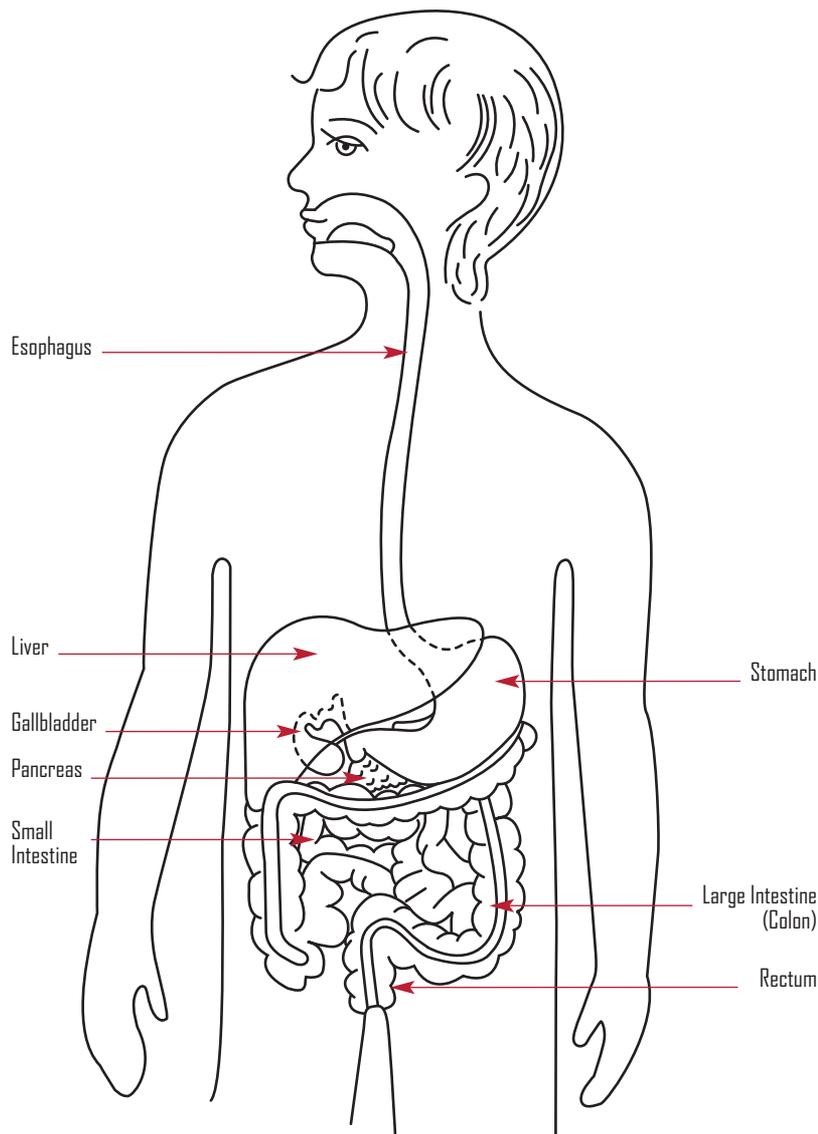
Digestion* takes place in the **gastrointestinal (GI) tract***. The GI tract is also called the digestive tract. The GI tract is basically a long tube that begins with the mouth and continues through the **esophagus***, stomach, small, and large **intestines***. (The small and large intestines together are about 25 feet long!) The GI tract ends at the **rectum*** and **anus***.

THE PANCREAS AND LIVER

Two other organs found in the **abdomen*** (belly) help with digestion: the **pancreas*** and the **liver***. The pancreas is an organ that sits in the upper abdomen behind the stomach. The pancreas produces **enzymes*** or special proteins that break down **fat*** and **protein*** in food. These enzymes include **lipase***, **protease***, and **amylase***. The enzymes pass into

the small intestine through a series of tubes. When there is food in the small intestine, the enzymes help break the food down so it can be absorbed and used by the body. The pancreas also produces **insulin*** that helps the body use **glucose***, a sugar that comes from the digestion of **carbohydrates***. Insulin is released into blood that passes through the pancreas.

The liver is an organ that sits in the upper right side of the abdomen. The **gallbladder*** is attached to the liver and helps store extra **bile*** fluid that is made by the liver. The liver and gallbladder are connected to the small intestine by a tube. The liver does many things for the body. Bile fluid is sent from the liver to the small intestine to help with digestion. The liver also helps with digestion of **nutrients*** in the blood that passes through the liver. The liver helps break down certain medicines in the blood and is a filter that helps clean the blood. The liver stores some nutrients for later use. Like the lungs, the liver has a lot of reserve function but it can fail if it is very damaged.



THE GASTROINTESTINAL (GI) TRACT

*See CF Words to Know Glossary.



THE STEPS IN DIGESTION

When food is swallowed it passes through the esophagus to the stomach. In the stomach, it mixes with stomach or **gastric fluid**.* Stomach fluid is made up of special acids that help break down food. The stomach is a muscle that also churns the food to help it mix with the stomach acid and break down into smaller pieces.

The food then passes into the small intestine. Most digestion takes place in the small intestine. The liver sends bile fluid to the small intestine to help with digestion. The pancreas sends special proteins called enzymes to digest fat and protein nutrients. The small intestine itself has enzymes that can break down some sugars and other nutrients. The small intestine breaks food down into small particles and chemicals that can be used by the body.

Absorption* is the other important thing that happens in the intestines, especially in the small intestine. When food is

digested it is broken down into water and tiny particles. These nutrients are the **calories*** of energy and materials the body's cells need to function. Nutrients pass through the lining of the small intestine into the bloodstream. This is called absorption. Nutrients are carried in the blood to all the cells of the body. They are used to keep the body's cells running well. They are also used to repair cells and for growth.

Undigested food travels on to the large intestine. Some water is absorbed in the large intestine or **colon**.* **Bacteria*** in the large intestine help break down the food as well. These good bacteria live in the intestines all the time and help digest food.

The leftover waste that cannot be used by the body makes up a **bowel movement*** or stool. The intestines have muscles that help squeeze and move the bowel movement down the tube until it passes out of the body through the rectum and anus.



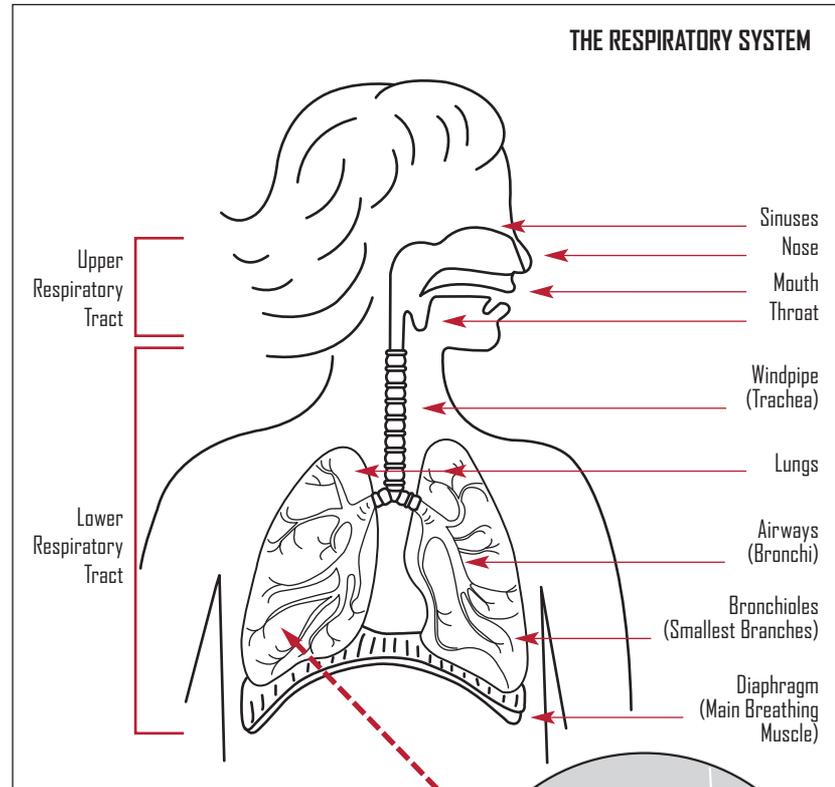
APPENDIX 4: CF FACTS – THE RESPIRATORY SYSTEM

Breathing is automatic. A special part of our brain controls our breathing without us even having to think about it. Each time we take a breath, air that passes in and out of the **lungs*** provides our bodies with oxygen that we need to live.

The **respiratory system*** contains the parts of the body responsible for breathing—the act of taking in oxygen and releasing **carbon dioxide***. The respiratory system is divided into the upper and lower respiratory tracts.

THE UPPER RESPIRATORY TRACT

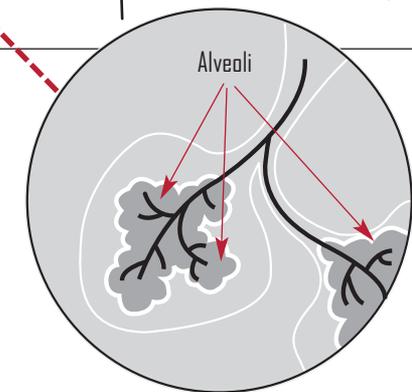
The **upper respiratory tract*** includes the nose, **sinuses***, mouth, and throat. The nose and sinuses help warm and filter the air we breathe. As the air passes through the nose, mouth and into the vocal cords in the throat, moisture is added to the air before it enters the **windpipe*** or **trachea***. (The windpipe is part of the **lower respiratory tract***.) The movement of air back and forth through the vocal cords is how we make sounds and talk.



THE LOWER RESPIRATORY TRACT

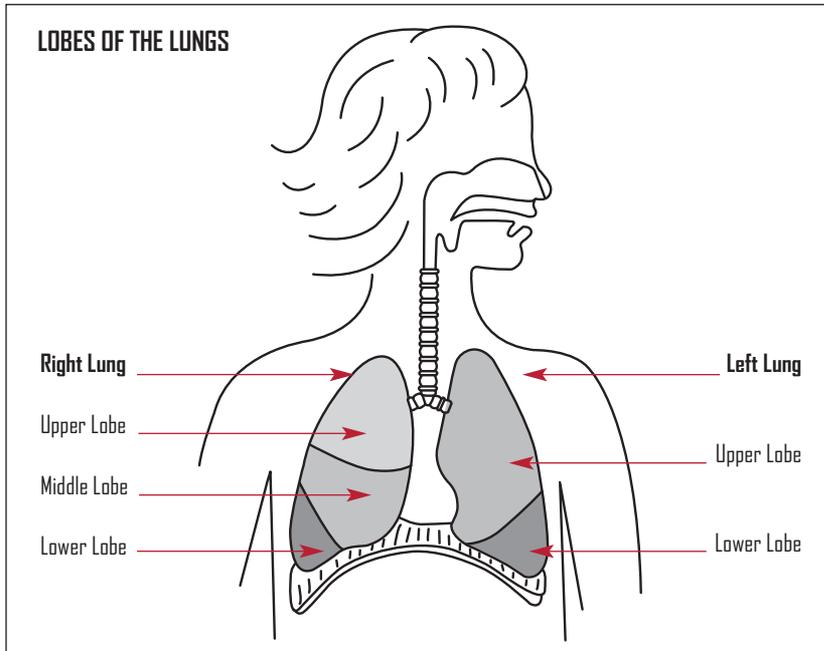
The lower respiratory tract includes the windpipe (trachea), airways, and lungs. The lungs and windpipe inside our chest are protected by the rib cage. When we breathe in, air flows down the trachea into our lungs. The trachea divides into two large airways called **bronchi***. Each of these tubes carries air to one of the two lungs. The airways divide like a tree in the lungs into smaller and smaller airways.

The smallest branches of the airways are called **bronchioles***. They carry the air deep into the lungs. The bronchioles end in tiny **air sacs*** called **alveoli***.



The alveoli are grape-like clusters of air sacs. There are millions of alveoli in the lungs. The alveoli give the lungs the look and texture of a sponge. The process of **gas exchange*** (breathing in oxygen and breathing out carbon dioxide) takes place in the alveoli.

*See CF Words to Know Glossary.



LOBES OF THE LUNGS

Each lung is made up of several sections called **lobes**.* The right lung has three lobes (upper, middle, and lower). The left lung has two lobes (upper and lower). The left lung is somewhat smaller because the heart also has to sit in the left side of the chest.

GAS EXCHANGE

The lungs provide oxygen for the whole body and get rid of carbon dioxide. Carbon dioxide is a gas that is part of the waste from our cells as they use oxygen. Our body needs to breathe out carbon dioxide to stay healthy. Oxygen is used for many functions in the cells. The air we breathe at sea level is made up of 21% oxygen. The air is thinner and contains less oxygen at high altitudes, such as in the mountains.

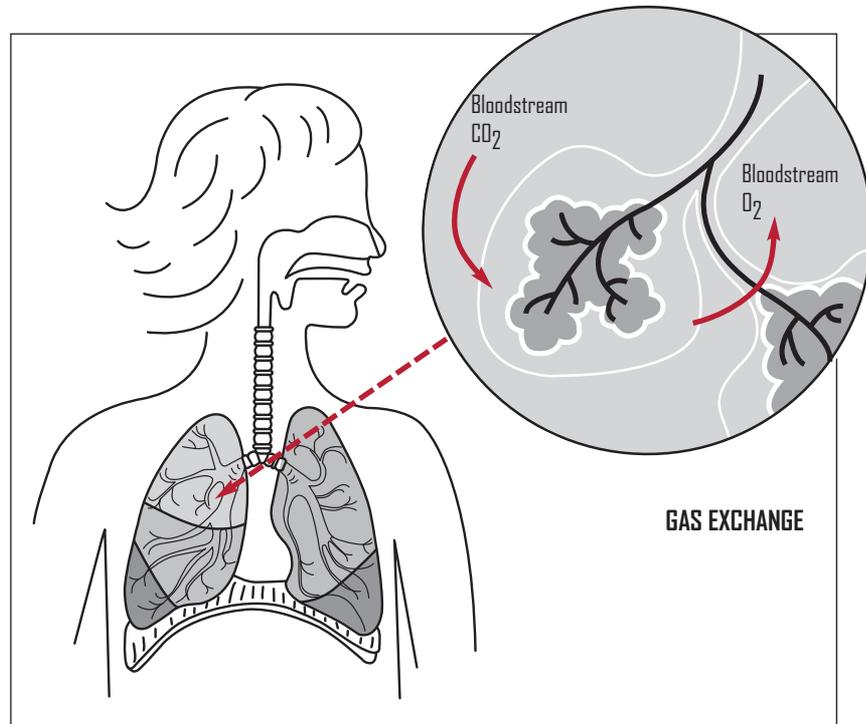
This exchange of carbon dioxide for oxygen takes place in the alveoli. The walls of the alveoli are very thin and are covered with tiny blood vessels. Oxygen passes through the walls of the

alveoli into the blood. At the same time, carbon dioxide moves from the blood into the air sacs. As we breathe out, carbon dioxide passes out of the alveoli into the bronchi and out of the lungs.

MUSCLES FOR BREATHING

To bring air into the lungs, the body uses several muscles. The main muscle used to breathe is the **diaphragm**.* This big muscle is just under the bottom of the lungs and separates the lungs from the **abdomen**.* When we breathe in, the diaphragm muscle pulls down. This creates suction in the chest that helps expand the lungs and draw air into the airways.

There are also muscles between the ribs and in the neck and the abdomen. These muscles are used more for coughing and when the lungs have to work harder than normal.





APPENDIX 5: WORKING WITH YOUR CF CENTER AND YOUR INSURANCE COMPANY

CYSTIC FIBROSIS SPECIALIZED CARE CENTERS

About 115 **cystic fibrosis*** (CF*) centers are accredited by the CF Foundation in the United States. CF centers meet or exceed medical care standards that the CF medical community feels are necessary to care for this complex and serious **chronic*** disease.

The health care professionals who work in CF centers specialize in treating CF. Going to a CF center gives families access to a team of people who know how to take care of CF and have the right care plan for a child with CF. CF centers can help parents and their children:

- Manage the disease while living as normal a life as possible
- Prevent **complications*** or problems from CF
- Identify problems early
- Treat complications effectively
- Teach children with CF how to care for themselves and become independent when they are adults

The CF Foundation not only accredits pediatric (children's) CF centers, but also accredits adult CF centers. People with CF usually transition to adult CF center care late in their teen years or as young adults. To find out more about the CF Foundation's CF Care Center Network visit their Web site at <http://www.cff.org>.

VISITING THE CF CENTER

The CF Foundation recommends that every person with CF be seen four times a year (quarterly) by a CF health care team. The quarterly visits are important because the CF health care team can review how well treatments are working and identify problems early. A person with CF should still do quarterly visits even if he or she feels fine. If you have problems getting to the CF center, you may want to ask the CF center **social worker*** for help with transportation.

ROUTINE HEALTH CARE

Most centers want you to have a **primary care provider*** for your child's routine health care including **immunizations*** (shots); well-child visits; common illnesses or problems, such as ear **infections*** and rashes; and sports or school annual physical exams. Your primary care provider might be a **pediatrician***, family doctor, **physician assistant***, or **pediatric nurse practitioner.***

YOUR CHILD'S PRIMARY CARE PROVIDER AND CF CARE

Typically your child's primary care provider does the routine health care (for example, school physicals or shots). But, if you live a long way from your CF center, you may need to have your primary care provider help you in an emergency or work together with the CF center to carry out a treatment plan in between your child's CF center checkups. If a CF problem does not respond well to treatment, your next step would be to call or visit the CF center. Talk with your CF health care team about how to work together with your child's primary care provider.

MEDICAL INSURANCE RULES

Medical insurance providers include HMOs (Health Maintenance Organizations); PPOs (Preferred Provider Organizations); private insurance; military insurance, such as CHAMPUS; and state programs, such as Medicaid or CHIP (Children's Health Insurance Plan). Each insurance program can have different ways of providing access to **specialists***, including your CF health care specialists.

Many plans require people to use only certain doctors or hospitals, usually called the plan's "network" or "preferred" providers. The CF center and its doctors may or may not belong to your plan's network. Some plans may not allow the person to go outside the plan's network for help, even if a person needs special care. In some plans, people can go outside the network for care as long as they get approval first. Such plans usually have doctors, nurses, or case managers who review a person's situation before granting approval.

Talk with your medical insurance representatives or case manager about the doctors in the network and how you can make sure that CF specialists see your child. Find out what your plan's requirements are for your child to be seen at the CF center. If your health care plan wants your child only seen by doctors in its network, find out how you can request that the CF center doctor be added to the network.

*See CF Words to Know Glossary.



Some health care plans may want you to see the CF center only once a year or when your child is having problems. Other plans may ask that you get approval before each CF center visit from your primary care provider. There are many possibilities. You may decide what your health plan requires is not in the best interest of your child. In that case, there are steps you can take.

- Work with your CF center to find a medical care plan that best matches the needs of your child and family.
- Don't be afraid to pursue matters with the insurance company and case manager until you feel comfortable that your child is getting the care he or she needs.

Keep in mind that you are your child's **advocate**.* You are the one to plead for and speak in support of your child getting good CF care. You do not have to take 'no' for an answer. You may need to be more assertive or forceful than you have been in other situations. Every insurance company and health plan has a formal appeals process. An appeal is a way of asking for a new review of a decision usually by a higher authority. This process may be difficult and time-consuming, but it can be worthwhile. Families may not receive everything they want for their children's CF care, but they are often able to reach an acceptable compromise with their insurance companies.

MANAGING PROBLEMS WITH HEALTH INSURANCE COVERAGE

Here are tips for managing your insurance coverage on a daily basis and when special problems arise.

- **Talk with a CF team member if you feel you are not getting what you need from your child's health care plan.** Team members may be able to tell you what has worked for other families in similar situations. The center can also be an advocate for your child's receiving the most appropriate care. You can learn more about health insurance coverage on the CF Foundation Web site: <http://www.cff.org>.
- **Know your health care plan's requirements.** Know about what is needed to get pre-approval for a clinic visit, hospitalization, or surgery. Know how many days ahead of time an approval is needed or when a claim must be filed. Bring your insurance card with you to clinic visits.
- **Get to know your child's insurance case manager.** Most insurance companies have case managers who can work with individuals and their families over time. Request a case manager if you have not yet been assigned one. Talk with your child's case manager to help him or her get to know your child's health problems and needs for CF care.
- **Be careful when considering a change in insurance coverage.** Plans may have a pre-existing condition clause. With a pre-existing clause, you could have a waiting period before your child's CF care is covered or there may be different charges because your child has a known chronic disease. Some states have laws to protect against this.
- **If your employer changes plans or you consider a change in insurance providers, see if your CF center doctor is in the network for the new plan.** Doctors in the network have agreed to accept the insurance plan's allowed amount for payment—non-network health care providers have not. If you use a non-network health care provider, you may be billed for charges over the insurance company's allowed amount in addition to your usual deductible and co-pay.
- **Check out whether there is any limit on how much a plan will pay.** Most have a limit on the amount of lifetime coverage for a person—this is called a cap. Compare insurance plans and what caps they may have. If you have questions or concerns, talk to a social worker or financial counselor for more help.
- **Do not let your child's health insurance expire.** Make sure you keep up with payments and renewals. Find out how often you have to renew your child's coverage even if you are in the state Medicaid plan that you do not pay for. A lapse in insurance coverage may make it more difficult to get coverage later.

**HEALTH CARE COSTS AND TESTING RULES**

Health care plans vary in the way they provide coverage for CF care. You may feel pressure to arrange your child's CF care in a certain way to get insurance coverage for the costs involved.

Some health plans may ask that any tests or **X-rays*** be done at a specific hospital or lab that is different from the one your CF center usually uses. Sometimes this works out without a problem. Sometimes it can be a hassle, but the results of the tests are still accurate and available to your CF health care team.

Sometimes the CF center, however, needs a certain kind or quality of test that the average lab cannot provide. Talk with your CF center about your health plan's requirements. One approach is to go with the health plan lab on a trial basis and let your CF center see if the results are acceptable. If there is a problem, you can talk with your health plan case manager or administrator. If a test, such as a **sputum culture***, requires a special lab to get the

best results, your CF health care team can help you talk with the health plan about why this special testing is needed. Ask the CF health care team for a letter about the test to send to your health plan.

Some medical plans have mail-order pharmacy services that are less expensive if you order 90-day supplies. If your co-payment will be less, ask your CF center if you can have mail-order prescriptions. The CF Services Pharmacy can provide information about patient assistance programs available from some drug companies. You can find out whether your insurance works with CF Services Pharmacy and how the costs compare by calling 1-800-541-4959 or by going to its Web site at <http://www.walgreens.com/topic/pharmacy/cystic-fibrosis-services.jsp>. You can also access it through the CF Foundation Web site at <http://www.cff.org>.

*See CF Words to Know Glossary.



APPENDIX G: TAKING YOUR CHILD'S TEMPERATURE

If your child seems warm to the touch, flushed, or you suspect he or she has an **infection***, use a thermometer to check for fever. Do not rely on your sense of touch. The doctor will want to know how high your child's fever is. There are different ways to take a child's temperature—some are more accurate than others. Often when parents go to the store, they are faced with many different types of thermometers. Knowing what different thermometers are for and what is most appropriate for your child's age will help you select a thermometer. After that, it is a matter of learning where to take your child's temperature.

USING DIGITAL OR ELECTRONIC THERMOMETERS

If you have not purchased a thermometer in a while, you will find that the glass thermometer your parents may have used has been replaced by the digital or electronic thermometer.

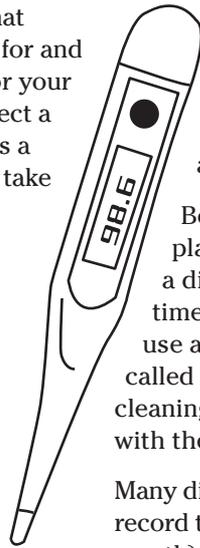
Note: The American Academy of Pediatrics recommends that parents not use glass thermometers with mercury because, if they break, there could be a risk of mercury poisoning. If you still have a mercury thermometer in your medicine cabinet, throw it out and replace it with a digital thermometer.

Note: New thermometers keep appearing on the market, but what will be described here are the more common digital thermometers.

Digital thermometers have small batteries that use electronic heat sensors to measure the body's temperature. The temperature is displayed in a window on the thermometer. Digital thermometers

are faster and easier to use than the older style thermometers. Digital thermometers usually don't cost much. You need to check your digital thermometer from time to time to make sure it is still working. The battery can run out. If your thermometer stops working, replace it with a new one.

Read the instructions on how to use the thermometer and turn it on before you use it. Most digital or electronic thermometers beep or flash when they are ready to read.



Be sure to use the disposable plastic covers that come with a digital thermometer. Each time you take a temperature, use a new plastic cover (also called a probe cover). Follow the cleaning instructions that come with the thermometer.

Many digital thermometers can record temperatures **orally*** (in the mouth), **rectally** (in the bottom), or **axillary** (under the arm).

ORAL, AXILLARY, OR RECTAL TEMPERATURE?

Here are some tips on where to take your child's temperature based on his or her age.

- **Infants—Birth to 3 months:** If your baby is less than 3 months old, the best way to take his or her temperature is by placing a thermometer under the arm or in the **rectum**.* The underarm or **axillary temperature*** is not as accurate as a **rectal temperature***, but if your baby or child is sleeping, you may be able to do an axillary temperature more easily. An axillary temperature above

99.4 degrees Fahrenheit (°F) means your child probably has a fever. But to make sure, take a rectal temperature. **The rectal temperature is the most accurate measure.** Check to see that you are using the right thermometer tip for a rectal temperature.

If you take a rectal temperature and your baby has a fever, call your doctor to have your baby seen right away.

There are oral electronic pacifier thermometers available now for infants, but ask your doctor before you buy one to find out how to use them and if they are accurate.

- **Children under 4 years old:** Older infants and toddlers who are less than 4 years old can have their temperatures taken either rectally (in the bottom) or axillary (under the arm). Again, the most accurate measure is a rectal temperature. Young children usually will not hold an oral thermometer in their mouths long enough for you to take a temperature. **Oral temperatures*** can be accurate, but they must be done correctly. There are electronic pacifier thermometers and ear thermometers available. These may not be as accurate. You should ask your doctor before buying one.
- **Children 4 years and older:** Children 4 years and older can have their temperature measured with an oral digital thermometer by placing it in the mouth under the tongue. You can also take an axillary temperature, but it may be less accurate. There are ear thermometers, but you should ask your doctor how accurate they are before buying.



HOW TO TAKE DIFFERENT KINDS OF TEMPERATURES

Follow the steps below for taking rectal, axillary, and oral temperatures. If you are unsure how to use your thermometer correctly, bring the thermometer to clinic with you and ask your child's primary care provider or the CF nurse.

Taking a Rectal Temperature

1. Clean the thermometer. Place a disposable plastic (probe) cover over the tip of the thermometer.
2. Put a water-soluble lubricant, such as K-Y® Brand Jelly, on the end of the plastic probe cover. Ask your **pharmacist***, doctor, or nurse what product to use.
3. Lay your child on his or her stomach and spread the buttocks gently so you can see the **anus***. If you prefer, you can place your child on his or her back. Then raise the legs as if you were changing a diaper. This position is harder with children who won't lie still.
4. Place the plastic-covered tip of the thermometer gently inside your child's anus. Be careful not to force the thermometer. If you put it in too far, you may hurt your child.
5. Keep the thermometer in place until it beeps or flashes that it is ready to be read. Make sure your child does not move or roll around.

NOTE TO PARENTS:

Parents sometimes panic when their child runs a very high fever. A temperature of 104°F or even 105°F is not rare among children under three when they are sick. Discuss what to do about a high fever with your child's doctor ahead of time.

6. Remove the thermometer and read it.
7. Throw out the disposable cover and clean the thermometer.

Taking an Axillary (Underarm) Temperature

1. Clean the thermometer. Place a plastic (probe) cover over the tip of the thermometer.
2. Make sure your child's underarm is dry and that there is no clothing between the arm and chest.
3. Carefully place the plastic-covered tip of the thermometer high up in the child's underarm. Make sure the tip is in the middle of the armpit, not sticking out the back or front.
4. Hold your child's arm against his or her body. Keep the thermometer under your child's arm until it beeps or flashes that it is ready to be read.
5. Remove and read the thermometer.
6. Throw out the disposable cover and clean the thermometer.

Taking an Oral Temperature

1. Clean the thermometer. Place a plastic (probe) cover over the tip of the thermometer.
2. Wait until your child has not had anything to eat or drink for 5 minutes.
3. Place the plastic-covered tip of the thermometer in your child's mouth, under the tongue.
4. Have your child close his or her lips gently to keep the thermometer in place.
5. Keep the digital or electronic thermometer in place until it beeps or flashes that it is ready to be read.
6. Remove the thermometer and read it.
7. Throw out the disposable cover and clean the thermometer.

REPORTING A FEVER

A temperature of 100.4 degrees Fahrenheit (°F) or higher is considered a fever.

Sometimes people leave out the decimal point when they read temperature numbers. When you talk to the nurse or doctor, it is important to say the word "point" with the numbers. For example, a temperature of 100.6 degrees Fahrenheit (°F) is read "100 point 6," not "100 and 6."

TREATING A FEVER

If your child is running a fever, there are many possible reasons. For example, your child may have a viral infection, such as a cold, an ear infection, or chickenpox. A **lung infection*** in CF does not always cause a fever.

Your child's primary care provider should give you instructions on how to treat a fever. You may want to write down your primary care provider's instructions on the worksheet "What to Do if My Child Has a Fever." (See sample worksheet below.) Ask your CF health care team for a worksheet or check the back pocket of the Cystic Fibrosis Family Education Program modules.

The worksheet is titled "What to Do if My Child Has a Fever" and includes the following sections:

- Header:** "What to Do if My Child Has a Fever" on the left and "Worksheet" on the right.
- Section 1:** "I will take my child's temperature _____ (date) _____ (time)." with a line for the date and time.
- Section 2:** "My child has a fever if the reading is more than _____ degrees Fahrenheit (°F)." with a line for the temperature.
- Section 3:** "If my child has a fever I will treat it by: _____" followed by four blank lines for notes.
- Section 4:** "I will call _____ if _____" followed by two blank lines for contact information.
- Footer:** "Copyright © 2005 Baylor College of Medicine" on the left and "Cystic Fibrosis Family Education Program" on the right.

*See CF Words to Know Glossary.



APPENDIX 7: KEEPING YOUR CHILD HEALTHY — ELIMINATING TOBACCO AND NICOTINE SMOKE EXPOSURE

Even those who do not smoke can be at risk for health problems when they breathe in air polluted with smoke from other people’s cigarettes, cigars, or pipes. This is called **secondhand smoke exposure**.* Exposure to vapors in the air from using **electronic nicotine delivery systems (ENDS)***, such as e-cigs, e-cigars, or e-hookahs, also is not healthy. A person does not need to smoke to be hurt by tobacco smoke or nicotine vapors. Just being around these can be harmful to your child’s health. There is no safe level of tobacco and nicotine exposure.

Smoking, vaping, or allowing it in your home, also gives kids a message that it is okay and can increase the risk of them becoming smokers, even if they have lung problems.

What makes tobacco smoke such a serious problem? Tobacco smoke:

- Has many harmful chemicals in it
- Stays in the air a long time, even after the cigarette, cigar, or pipe is put out
- Can move from room to room
- Can hurt your child’s **lungs***

While less is known about vapors produced by ENDS devices, other chemicals (along with nicotine) are released in the air that are not healthy to breathe.

It is now known that chemicals and particles in tobacco smoke and ENDS vapors can land on surfaces and get back into the air. This is called **thirdhand smoke exposure*** and is another reason to keep smoke out of your home and places where your child spends time.

RISKS OF SECONDHAND SMOKE EXPOSURE

Healthy children who grow up in homes where someone smokes are more likely to have:

- Colds
- Ear **infections***
- **Bronchitis***
- **Pneumonia***
- Reduced **lung function***
- Lung cancer later in life (even if they never smoke themselves)

Children who already have a lung disease, such as **cystic fibrosis*** (CF*) or **asthma***, are at even greater risk for health problems when they are around tobacco smoke. **Research*** has shown that children with CF who are regularly around tobacco smoke have a greater decline in lung function, and their disease worsens or progresses more than those who are not exposed. Lung damage from secondhand smoke exposure can happen silently for a long time. The damage can be there even if your child does not seem to have any **symptoms**.*

WHAT YOU CAN DO

There are two things you can do to limit your child’s exposure to tobacco smoke:

1. Make your home a smoke-free zone
2. Limit the amount of tobacco smoke-polluted air your child is exposed to

Make Your Home Smoke-Free

Take the smoke-free pledge and eliminate smoking in your child’s home. First, see who smokes in your home. Who are the smokers in your child’s world?

- ✓ You
- ✓ Grandparents or other members of the family
- ✓ Friends
- ✓ Babysitters

Decide as a family to enforce a no-smoking policy.

Eliminating smoking and smoke exposure is ideal. Having people smoke outside doesn’t solve all the problems, but smoking outdoors is much better than smoking indoors. Even though a person may smoke outside, you can still smell smoke on his or her clothes.

If smoking outside is not possible, make one room the only place people can smoke inside. Choose a room with good ventilation—a room where you can open the windows to let in fresh air. Air cleaners do not filter out all the toxins in tobacco smoke or nicotine vapors. Ceiling fans just spread smoke around. You have to be able to bring in fresh air. Do not allow smoking in rooms where your child sleeps or plays.



Limiting or Eliminating Your Child's Secondhand Smoke Exposure

Here are some ways to limit or avoid secondhand smoke:

- Sit in the *no smoking* section in public places and restaurants.
- Support efforts in your community to make smoking a problem of the past.
- Let other smokers who may be around your child know how important it is to keep tobacco smoke out of your child's lungs. Tell them it is your child's doctor's order. You may want to give other people who smoke a copy of a "Dear Smoker" letter or a prescription for clean air from your CF health care team. Ask your CF health care team for a "Dear Smoker" letter or check the back pocket of the CF Family Education Program modules.
- Smokers may find that by making changes in where they smoke, they end up smoking less. Being able to cut back to help protect others is a great first step to quitting smoking as well!

QUITTING SMOKING

If you smoke, the best thing you can do for your child, your family, and yourself is to quit smoking. You should also encourage others who smoke to quit. Quitting smoking will have definite health benefits for you, no matter how long you have been a smoker. It also can save you money.

Quitting smoking can be very hard, but it's not impossible. There is no "magic bullet" to quit smoking, but there are many resources and support systems you can use to help you:

1. Talk to your doctor about using medicine to help with nicotine dependence. You may want to try nicotine replacement products you can buy without a prescription.
2. Enlist a friend or family member for support.
3. Believe in yourself. You CAN quit! Most people try 3 to 4 times to quit before they are finally successful—so keep trying.

Electronic Nicotine Delivery System (ENDS) Devices

E-cigs and other devices turn nicotine liquids into a vapor that can be inhaled. The nicotine liquids have other chemicals in them (including flavorings) that can be harmful to your health and those who are exposed to the vapors (smoke).

Some people will use these devices as a substitute for tobacco. Keep in mind that there are still risks and you should not be exposing your child to these vapors.

Resources to Help You Quit

Check out these self-help resources on the following Web sites:

- <http://www.smokefree.gov>
- http://www.cdc.gov/tobacco/quit_smoking (Centers for Disease Control and Prevention)
- <http://www.lung.org/stop-smoking/> (American Lung Association)

All of these Web sites have free material you can read and download to use and share.

You can also call and get free expert help:

- The National Cancer Institute free quit support line at 1-877-44U-QUIT. For help within your state, call 1-800-QUIT NOW.
- You can also get information and advice from counselors through instant text messaging by using NCI's LiveHelp service on their Web site: <http://smokefree.gov/talk-to-an-expert>.

Help your child demand clean air to breathe. Show your child the value of being a nonsmoker. Never give up on the idea that all the smokers in your child's life will one day quit, letting everyone breathe easier.



APPENDIX 8: HOW TO DO BREATHING TREATMENTS WITH YOUR INFANT OR TODDLER

When infants and young children do **breathing** (or respiratory) **treatments**,* they **inhale*** medicines directly into their **lungs**.* The **cystic fibrosis*** (CF*) doctor prescribes several types of inhaled medicine to treat CF. Some medicine, such as Pulmozyme®, helps loosen **mucus**.* Some medicines, including albuterol or levalbuterol, relax the **airway*** muscles to relieve **bronchospasm**.* Inhaled **corticosteroids*** and other medicines treat **inflammation***. Inhaled **antibiotics***, such as TOBI®, treat **infection**.* Your CF health care team will help you decide which inhaled medicines your child should take and how these can best be given to your child.

Infants and toddlers inhale medicines either through a **nebulizer*** as an **aerosol*** mist or with a **metered dose inhaler*** (MDI) using a valved **holding chamber*** and mask. The steps on the next page will help you use the equipment correctly so that your infant or child is inhaling as much medicine as possible.

Before you follow the tips for giving breathing treatments, make sure you have your child sit up, if possible. Sitting up allows your child to take deeper breaths. You may find it easiest to hold your child in your lap, facing forward. Some babies are calmer if they have a blanket wrapped around them and are held snug during the treatment.

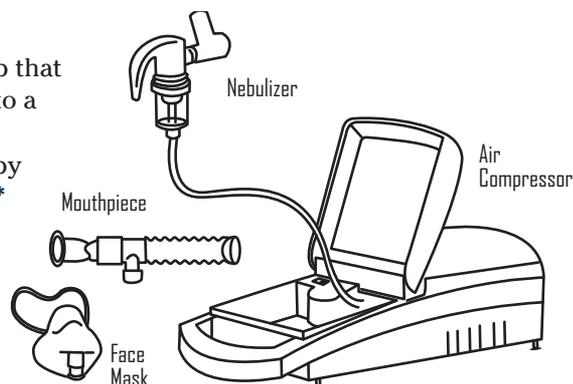


NEBULIZER AND MASK

A nebulizer is a plastic cup that turns a liquid medicine into a mist. Air is blown into the nebulizer through tubing by a medical **air compressor*** machine. The infant or young child inhales the mist through a **face mask***, or if the child is older, he or she uses a **mouthpiece**.* Even if a baby or young child can hold a mouthpiece like a straw, it is still recommended that a young child inhale medicine through a face mask. Young children will be able to inhale more medicine through a face mask because they breathe more through their noses than older children do. Usually when children are 5 years old, they can begin to use a mouthpiece. Ask your CF doctor or **respiratory therapist*** to help you decide when your child is ready to use a mouthpiece.

Some medicines used in a nebulizer come in a pre-mixed **vial*** that you just pour into the cup. Other medicines have to be measured. Make sure you know how to measure the liquid medicine correctly. Always check the parts of the nebulizer to make sure there are no cracks or leaks.

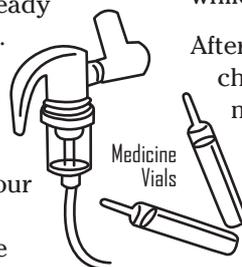
Some medicines require a specific type of nebulizer. Different nebulizers are designed to make different **particle*** sizes in a mist. The doctor, respiratory therapist, or **pharmacist*** can help make sure you have the right nebulizer.



Since a nebulizer treatment takes from 10 to 15 minutes, and children need to wear a mask, they may cooperate better if they are distracted. For your infant, you may want to try a mobile or music. If you have a toddler, you might sing songs or let him or her watch a special DVD or TV show during the treatment. Give your child a choice: “Do you want to watch *Sesame Street* or read a story while we do your treatment?”

After you place the mask on your child so that it covers his or her nose and mouth, you will need to hold or stay with your child the whole time. To avoid any interruptions, take care of your other children, phone calls, food on the stove, and other tasks before you start the treatment. New types of machines are being designed that may shorten nebulizer treatments, but for now, plan on at least 15 minutes for every treatment.

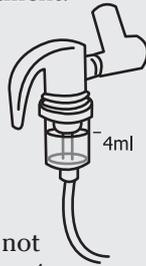
Note: If your child’s breathing treatment takes longer than 15 or 20 minutes, there may be something wrong with the air compressor or the nebulizer. Talk to your CF health care team if you think it is not working properly.





STEPS: Using a Nebulizer with Air Compressor

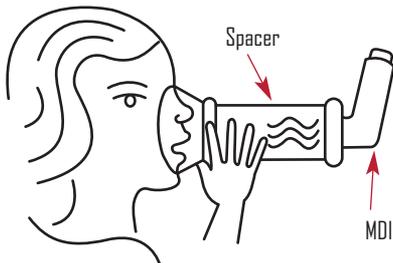
1. Remember to wash your hands before giving a treatment.
2. Obtain a unit **dose*** of medicine. If you need to give more than one medicine be sure that it is okay to mix the medicines. The nebulizer cup does not hold much more than 4 ml of medicine so you may have to give separate treatments. If you have to measure or mix a medicine solution, get instructions from the pharmacist or CF team. You should try to have 3 to 4 ml in the cup to get a good mist.



3. Set up the air compressor and attach the tubing. Be sure that you do not put the air compressor on a soft surface, such as a bed or couch, where the air openings and ventilation slots may be blocked.
4. Put together the nebulizer and connect it to the tubing.
5. Remove the top of the nebulizer and pour in the medicine. Replace the top of the nebulizer cup, being careful not to spill any medicine.
6. Attach the mask to the nebulizer cup. Place the mask over your child's nose and mouth. You can hold the mask or use the elastic strap around your child's head to secure it.

- The mask should feel comfortable, yet snug, so no air leaks out.
7. Have your child sit up or use pillows to prop up your child. You may want to hold a young child in your lap.
 8. Turn on the air compressor. Encourage your child to take slow, deep breaths.
 9. Stay with your child during the treatment. Continue the treatment until the mist stops and the medicine is gone from the nebulizer.
 10. Remove the mask from your child. Clean the mask and nebulizer as instructed.

METERED DOSE INHALER (MDI) AND VALVED HOLDING CHAMBER (SPACER)



A metered dose inhaler or MDI is a metal canister with a valve that releases a pre-measured puff of medicine when it is pressed down. (The MDI is also called an **inhaler*** or **puffer***.) The medicine comes out as a quick puff. It's best if you use a **spacer*** with an MDI to help hold the medicine so that your child can inhale it more slowly and deeply. Your CF health care team can show you how to use your child's spacer correctly.

STEPS: Using an MDI with Spacer

1. Remember to wash your hands before giving a treatment.
2. Shake the metered dose inhaler (MDI) and remove the cap.
3. Put the MDI mouthpiece into the spacer opening with the canister pointing up.
4. Place the mask snugly onto your child's mouth and nose. It must make a good seal so the medicine does not leak out.
5. Spray one puff of medicine into the spacer.
6. Have your child take four to six normal breaths with the mask in place. Some spacers have flaps you can watch move back and forth as your child breathes in and out.
7. Remove the mask from your child's face. To give another puff, wait 30 seconds, then repeat steps three through five.
8. Clean the spacer as instructed.

You can ask your pharmacist or CF health care team how to tell when the MDI needs to be replaced. Check the number of doses (**inhalations***) that a canister has in it. How long a canister lasts depends on how many inhalations (puffs) you use per day. You cannot tell if a canister is empty by shaking it because sometimes you can still hear the propellant when the medicine is gone. You should *not* float the MDI in water to see if it is empty. Putting the canister in water could clog the valve.

*See CF Words to Know Glossary.



CLEANING INSTRUCTIONS FOR RESPIRATORY EQUIPMENT

Germs*, including **bacteria*** and mold, can get in the equipment. You should clean equipment, including nebulizers and spacers, after each treatment. The plastic tubing that carries the air to the nebulizer does not require cleaning unless it is dirty on the outside. The tubing can be wiped off, but do not try to wash the inside. If it looks dirty on the inside, it should be replaced. You can wipe the air compressor on the outside. The filters on the inside should be changed according to the manufacturer's instructions. You can learn more about CF germs and why it's important to clean and disinfect respiratory equipment on the CF Foundation Web site (<http://www.cff.org>).

STEPS: Cleaning Respiratory Equipment

1. Clean your child's respiratory equipment after every treatment. Take apart the pieces of the nebulizer. Remove the back piece from the spacer only. Do not remove the face mask or valves.
2. Clean all parts with warm water and a mild dishwashing liquid.
 - Fill a bowl or sink with warm water and add a mild, clear detergent.
 - Submerge the spacer and nebulizer parts, and then swish back and forth gently.
3. Rinse by submerging in clean water.
4. Shake off the excess water.
5. Disinfect to destroy germs using heat method or a disinfecting solution described on this page under **Disinfect the Equipment**. Your CF health care team can help you find the best way to disinfect your equipment. Some equipment may require special care or specific methods for cleaning.
6. If a solution is used, rinse after disinfecting with sterile water (you can get this from a medical supply company or make it by boiling water for 5 minutes and then cooling it).
7. Let the equipment parts air dry on a clean towel or cloth.

Note: Some equipment can be damaged by using heat disinfecting methods. Routinely check the equipment for damage. The equipment does wear out with time. Replace cracked or broken parts. You may want to have two sets so you can have one clean at all times. Call your CF health care team if you need a prescription to replace equipment. Make sure you have the instructions for the type of equipment that you use.

Disinfect the Equipment

Disinfecting

Any of the following methods using heat to disinfect respiratory equipment can be used **IF** it does not damage the equipment. Your CF health care team can help you find out if you can use any of these with your equipment.

Heat disinfecting options:

- Using an electric steam sterilizer, (e.g., a baby bottle sterilizer).
- Placing in boiling water and boil for 5 minutes.
- Placing in water in a microwave-safe bowl or bag and microwave for 5 minutes.
- Using a dishwasher if the water is hotter than 158 degrees for 30 minutes. Check to see if your dishwasher has a "Sanitize" setting. (For more information, go to: http://www.nsf.org/consumer/newsroom/fact_safer_dishwasher_sap)

Disinfecting Solutions

Any of the following solutions can be used to disinfect respiratory equipment. You should make a fresh batch of any of these solutions each time you clean the equipment. **Do not leave disinfectant solutions in reach of children.** Remember, these chemicals could be dangerous. Pour the solutions out right after you finish cleaning. Remember to rinse with sterile water after disinfecting. Both of these solutions can be purchased without a prescription.

Disinfecting solutions and soaking times:

70% isopropyl alcohol	5 minutes
3% hydrogen peroxide	30 minutes



HELPING YOUR CHILD ADJUST TO THE FACE MASK

Toddlers and young children may resist using a spacer or nebulizer mask at first. They will adjust, however, to using the mask within several days if caregivers are consistent in using the face mask. Keep the face mask on until your child, whether crying or not, has finished the treatment. Your child will inhale more medicine if he or she is not crying. Use the suggestions in the shaded box to help your child adjust to the mask and treatments.



Remember: Even if young children can put a mouthpiece in their mouth, they will not inhale as much medicine as they would if they used a face mask.

STEPS: Helping Your Child Cooperate with Treatments

Follow these steps each time you do a treatment:

1. Tell your child it is time to take his or her medicine.
2. Ask your child if he or she wants to help hold the face mask or if you should do it. (If your child does help, make sure the mask is still held snugly to his or her face.)
3. Do not spend time trying to convince your child to put the mask on or that it will not hurt him or her. Negotiation will just delay the process. Make the decision on who holds the face mask within 30 seconds. Once you decide it is time, give the puffs even if your child is not cooperative at first.
4. After you have given your child the puffs, reinforce your child's efforts by saying, "Good job!" or "You're learning to take your medicine."

If you follow these steps the same way for each treatment, it will help your child learn to work with you to take the medicine in a short period of time.

NOTE TO PARENTS:

If your child does not begin to cooperate within a week or two, talk to your health care provider about a referral to a child **psychologist*** or other behavioral specialist.

For more information about **respiratory treatments*** and inhaled medicines, see the CF FEP module *Managing Lung and Other Respiratory Problems*, or go to the CF Foundation Web site (<http://www.cff.org>), or call 1-800-FIGHT-CF.

*See CF Words to Know Glossary.



APPENDIX 9: GIVING YOUR CHILD MEDICINE

You will probably be giving your child with **cystic fibrosis*** (CF*) medicine every day. How you give the medicine depends on the type of medicine and the age of your child. Here are some hints to help you.

THE “5 RIGHTS” FOR GIVING MEDICINE

Make sure you know the right way to give medicine to your child. One way to double-check is to go over the *5 Rights*:

1. **Right medicine?** Is it the drug that the doctor prescribed?
2. **Right form?** Is it a liquid, pill, or a capsule?
3. **Right amount?** How much should your child take?
4. **Right route?** Is this a medicine to swallow or breathe in?
5. **Right time?** Is this the time your child should take it?

WHAT TO ASK YOUR DOCTOR OR PHARMACIST

Do I give the medicine with or without food? If your child is supposed to swallow the medicine, find out if it should be given with food or on an empty stomach. With some medicines it does not matter. Be sure to check with your doctor or **pharmacist*** to find out what foods are okay to mix with a given medicine.

Will the medicine interact with any other medicines my child is taking? The pharmacist or doctor can tell you if certain medicines do not mix well or could interact with each other.

What if my child spits out the medicine? Ask the doctor ahead of time what to do if your child spits out or throws up the medicine.

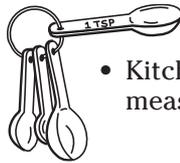
What about side effects? Know what **side effects*** to watch for if you are giving your child a new medicine.

FORMS OF MEDICINES

Liquid Medicines

Babies and toddlers usually need liquid medicine.

Measure liquid medicine with a:



- Kitchen measuring spoon

- Special medicine spoon



- **Medicine syringe***

You can get the medicine spoon and syringe at a drugstore. You should not use the kind of teaspoon you eat with because teaspoons for eating can vary in size and may not be an accurate measure.

The medicine syringe does NOT have a needle, but is actually a tube with markings on the side to measure medicine. The tube with a plunger allows you to squirt the medicine into your child’s mouth. To use a medicine syringe, squirt the medicine inside the cheek. If you squirt it directly into the back of the throat, it could make your child choke. A baby may suck on the syringe like a nipple.

You can also try putting a small amount of medicine into a nipple and hold the nipple while your baby sucks the medicine.

Do not add medicine to a full bottle because your baby might not drink all of it and he or she will not get the full dose of medicine.

Sprinkle Medicines

Some medicines (such as **pancreatic enzymes***) come as beads in capsules or packets that can be opened and sprinkled on food. Be sure to check with your doctor or pharmacist to find out what food is okay to use with a medicine.

Pills or Capsules

By six or seven years old, most children can learn to swallow pills and capsules. See the module *Working with Your Child* to find ways to help your child learn to swallow pills.

TAKING MEDICINE TO DAY CARE OR SCHOOL

In many states it is against the law for day-care or school staff to give your child medicine unless the medicine is in a labeled bottle from the pharmacy. Ask the pharmacist to split the prescription into two labeled bottles: one for home, one for day care. That way you will not wind up at home on the weekend with your child’s **antibiotic*** locked up in a day-care or school refrigerator!

NOTE TO PARENTS:

If you have questions about any medicine, ask your CF health care team for information.

Let your CF health care team know if you think your child is having side effects from a medicine, or if you are having trouble giving your child the medicine.



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