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Introduction

May 2010

Dear Families,

Hearing the words "Your child has Cystic Fibrosis" for the first time can be devastating. Your world, your hopes, your plans are suddenly topsy-turvy. You may wonder, "What can I do now? How will I get through this? How will I help my child, my family, my marriage?" The list of questions is endless. Yet, so are the possibilities and outcomes.

This handbook was created by families that have all asked these very questions. We have, and continue to, walk in your shoes, down the same path, with the same goal. The parents and professionals on the CF Family Council have created this manual to help you through the many facets and phases of Cystic Fibrosis. It is our hope that this book will be a tool that empowers you to be a great advocate for your child. On this path, you will meet many strong and giving families and professionals. We are all part of a support network for you and your child now.
Please seek our help.

This handbook is one gesture of assistance. We hope you'll use this as a resource to discover and find strategies for working with professionals, organizing your child's routines, preparing for changes in your child's life, accessing financial aid, finding social and emotional support, and how to support further the research and progress towards a cure for Cystic Fibrosis.

Most sincerely,

Doernbecher Cystic Fibrosis Family Council

1 What is Cystic Fibrosis?

In this chapter:

- What CF is not
- How a diagnosis is made
- General treatment suggestions

Chapter 1
Cystic Fibrosis Synopsis and Overview

What is Cystic Fibrosis?
Cystic fibrosis (CF) is an inherited disorder that affects the movement of salt and water into and out of the body's cells. The problem affects many parts of the body, but is especially damaging to the lungs and digestive system.

CF affects ***exocrine glands,*** which normally produce thin, watery secretions like mucus, tears, and sweat. With CF, mucus becomes thick and sticky, which can clog tubes and passageways.

In the lungs, small airways get plugged with thick mucus, which leads to disease and damage.

In the pancreas, a part of the digestive system, the blockage from the thick mucus causes poor absorption of food, which can lead to malnutrition and poor growth.

Cystic Fibrosis is NOT:

Contagious
CF is inherited and present at birth. You cannot "catch" CF.

Because of something we did wrong
Nothing you did during pregnancy causes CF.

Going to cause brain damage or learning problems
CF does not affect the brain or nervous system

Curable
At this time, there is no cure for CF, but there are excellent treatments available which allow children with CF to grow up and lead active, full lives.

How is CF diagnosed?
With CF, ***chloride*** levels are elevated in sweat, which can be measured by a ***sweat chloride test.*** This is the most common method of making a diagnosis. Another way to diagnose CF is with a blood test, which looks for the genetic abnormalities which cause the disease.

In Oregon and Washington, all newborns are screened for CF. Newborn screening is not a final diagnosis. If the screening test is positive, the child is sent to the OHSU CF Clinic for a sweat chloride test and further evaluation.

Doctors may test children for CF if they find certain symptoms, such as poor growth, lung problems, or digestive difficulties. The severity of CF cannot be determined by the sweat chloride or blood tests.

How did our child get Cystic Fibrosis?
CF is an inherited condition, present from conception. Everyone has two genes which determine whether or not they have CF. Both genes need to be abnormal for a diagnosis of Cystic Fibrosis.

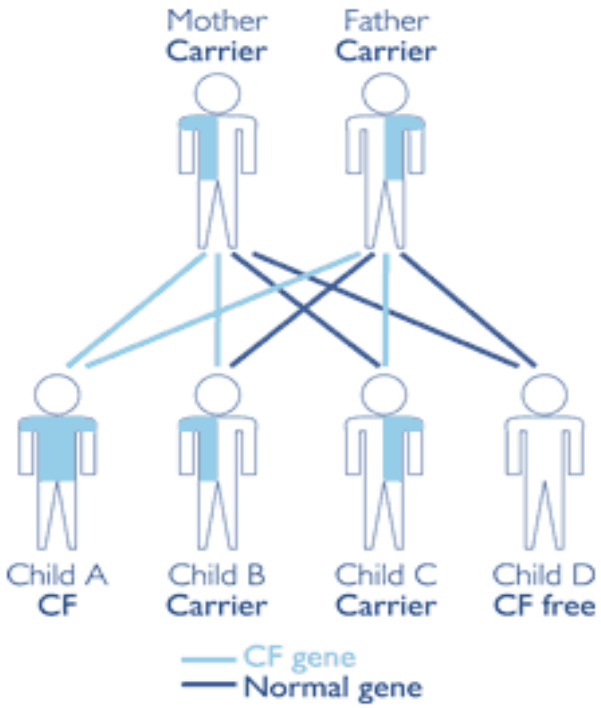
Individuals who have one normal and one abnormal CF gene are called ***carriers***. They do not have CF, but they can pass their abnormal genes to their children. To have CF, a child must inherit two abnormal genes, one from each carrier parent.

Every time two carrier parents conceive children, there is a 25% chance their child will have CF.

~ 25% chance (1 in 4): Cystic fibrosis

~ 50% chance (2 in 4): Child will inherit one gene and become a carrier like their parents

~ 25% chance (1 in 4): Child will inherit two normal genes and not have CF or be a carrier



There is no way to predict or change this outcome. Having one child with CF doesn't mean you can't have other children who will *also* have CF. Some families have several children with CF; some do not.

Does this mean we should have our other children tested for CF?
We recommend testing your child's siblings and half-siblings for Cystic Fibrosis, even if there are no signs or symptoms. CF can be present without obvious signs, even for many years. It's better to test early and get children into routine CF care if they are affected.

What are the signs and symptoms of CF?
Symptoms of CF can vary greatly from person to person. Your child may have some, all, or none of the following:

- Abnormal stools, including: diarrhea, foul-smelling, or large
- Greasy or oily bowel movements
- Poor weight gain and growth
- Excessive gas or stomach bloating
- Cough
- Frequent lung infections
- Chronic nasal or sinus infections

What causes our child to have abnormal stools?
Abnormal bowel movements are caused by a problem with the pancreas, called ***pancreatic insufficiency.*** Normally, the pancreas secretes enzymes which help our bodies digest food. For CF patients, tiny ducts within the pancreas are blocked with thick mucus which prevent the enzymes from reaching the digestive tract. The body is then unable to break down the food and digest it. Instead of absorbing the nutrients into the body, the food passes straight through the intestines. This causes stool to be bulky, greasy, and soft. Excess gas forms and causes abdominal bloating and painful cramping.

Why does our child get lung infections?
In the lungs, the thick mucus makes a sticky layer that lines the airway tubes. This sticky mucus acts like flypaper and traps bacteria that enter the lungs. Once bacteria ***colonize*** the lungs, they live there forever and cause damage from ***inflammation***. This is because the body tries hard to fight the bacteria, but it's a losing battle; the bacteria will always continue living in the lungs. This long-term fight slowly damages the lungs over time —over many years. This is why CF is called a ***progressive*** disease. The damage continues and is not reversible.

This sounds scary. What can we do to treat our child?
Being diagnosed with CF is a frightening and stressful time. Your CF care team provides education to help you effectively fight this disease with medicines and therapies. This may include different treatments, including medicine to help digestion and promote good growth, and treatments to help minimize the damage to the lungs. Therapy is tailored specifically to your child and their unique health care needs.

"*When Matthew was 11 years old, he told me he wanted to go out for track at his school. Usually at his grade level the students choose which events they would like to compete in. Matthew informed me he wanted to do the standing long ump, a softball throw, 200 meter relay, and the 400 meter relay. I was a little concerned that Matthew wanted to do two running events. Matthew not only has Cystic Fibrosis; he also has asthma and it was spring when allergies were in full swing. However, we decided a long time ago not to hold Matthew back from participating in life because of his disease(s) so I kept my reservations to myself and encouraged him all the way.*

Then the big day came. Matthew did the standing long jump and did it quite well—winning second place. Next was the 200 meter relay which also went well and it didn't seem to slow him down at all. The third event was the softball throw and I thought we were done for a while because the 400 meter relay was still a few events away. There was an announcement for the 400 meter race and to my surprise, Matthew got up and headed for the staging area. I called to him and said, "This is not your event, it is the race not the relay."

He then stated back to me, "Mom, I'm not in the 400 meter relay, I am running the race". Needless to say I was taken aback and for a split second I thought I should stop limit's way too far for him to run, but he was already off to join the "healthy" kids he would be competing against. So I sat with my eyes locked on my boy,my hands grasping his Albuterol inhaler ready to rush to his side if he needed me. When the starting gun sounded, Matthew took off with the rest of the group instead of falling back like I feared, my son started to pull ahead and by the last leg of this very long race he was in the lead.

As I watched with pride, Matthew —my 11 year old boy who suffers from not one, but two lung diseases—took first place that day in the 400 meter race, beating all eight of his peers. He went on to take sixth place in the district race later that month.

Matthew, now 13 years old, continues to inspire me and everyone around him. He lives each moment to its fullest, and never lets Cystic Fibrosis dictate what he can and cannot do with his life."

2 CF Clinic Providers

In this chapter, introductions to:

- Pulmonologists
- Nurses
- Dietician
- Social Workers
- Administrative Support

Chapter 2
CF Clinic Providers

Mike Powers, MD
Pulmonologist
Chief, Pediatric Pulmonary Division

Dr. Powers received his medical degree from OHSU in 1985. He continued his medical education at OHSU by completing his Pediatric residency and fellowship in Pediatric Pulmonary. He received his board certification in Pediatrics in 1989 and in Pediatric Pulmonary in 1992. Dr. Powers is an associate professor and his special interests include pediatric pulmonary infections, flexible bronchoscopy, growth factors, and lung injury. When he's not at the CF Clinic, he can usually be found working long hours in his research lab, and also enjoys playing golf, star-gazing, and singing with his rock 'n' roll group, The Moderator Band.

Michael Wall, MD
Pulmonologist
CF Clinic Director

Dr. Wall graduated from the University of North Carolina at Chapel Hill Medical School and completed his residency and fellowship in Boston, Massachusetts. He moved to Oregon in 1978 and became the Chief of the Pediatric Pulmonary Division and Director of the OHSU Cystic Fibrosis Center. He is a Professor in the Department of Pediatrics and is board-certified in pediatrics and pediatric pulmonology. He is the principal investigator for a number of Cystic Fibrosis drug development trials currently underway. When he's not working, he enjoys playing golf, sailing, and travel.

Holger Link, MD
Pulmonologist

Dr. Link graduated from Georg-August University, Gottingen Medical School, Germany in 1992. His internship and residency training was in England and Germany followed by a residency and fellowship in the United States. He is board-certified in pediatrics, pediatric pulmonology, and sleep medicine, and has been an assistant professor at OHSU since 2001. Dr. Link specializes in the treatment and care of kids with CF and asthma and has a strong interest in patient education. He also keeps busy as a sleep medicine specialist, working with kids who have sleep and circadian rhythm disorders. Dr. Link enjoys spending time with his family and loves to walk.

Kelvin D. MacDonald, MD RRT
Pulmonologist

Dr. MacDonald started his career as a respiratory therapist, eventually becoming a Clinical Research Coordinator in Los Angeles where he conducted clinical and bench trials related to pediatric respiratory care. He is certified in both general and pediatric respiratory care and has a Bachelors degree in Applied Science with a concentration in Respiratory Care from Thomas Edison State College. Seeking more challenges, Dr. MacDonald completed medical school at the University of Saint Eustatius, graduating with a Clinical Achievement award for obtaining honors in all clinical rotations. In 2004, he completed a pediatric residency at the Children's Hospital Medical Center of Akron, receiving awards for intensive care and pulmonology excellence. Dr. MacDonald then completed a fellowship in Pediatric Pulmonology at the Johns Hopkins University in Baltimore, Maryland and joined the faculty where he was actively involved in the CF Translational Research Center and served as the director of Nasal Potential Difference Laboratory. Dr. MacDonald has received research support and awards from the National Institutes of Health, Cystic Fibrosis Foundation, Thrasher Research Foundation, and the National Institutes of Child Health and Development. *"I am excited to be at OHSU, bringing my research and clinical efforts to such a wonderful, supportive environment in the beautiful Pacific Northwest"*. When he is not busy working on a cure for CF, Kelvin enjoys playing with his son, golf, and Pinot Noir.

Ben McCullar, RN
Nurse Coordinator

Ben graduated from the OHSU School of Nursing in 1989 and has worked at OHSU ever since. He began on an inpatient internal medicine unit, working with adult patients with a multitude of issues, including pneumonia, Cystic Fibrosis, diabetes, liver disease, and soft-tissue infections. He also moonlighted as a home health RN for CF patients and worked part-time as a CF Nurse Research Coordinator, eventually joining the pediatric pulmonary division full-time in 2003. He enjoys teaching students and families, and has a strong interest in making the experience of coming to the hospital and clinics as easy as possible for families with CF. He is married to Amy, also an RN at OHSU, and is a confirmed "serial hobbyist;" his current favorites being photography, playing guitar, hiking, bicycling, and world travel.

Emily Somervell, MSW
Social Worker

After receiving a BS in Psychology from Santa Clara University in 1999 Emily moved to Portland and began working at Morrison Child and Family Services Edgefield residential center. She worked with children with mental health challenges for three years. In 2002, she left Edgefield Children's Center to help bring Big Brothers Big Sisters back to the Portland Metropolitan area. In 2005, she received her MSW from Portland State University. During graduate school, she gained experience working in and elementary school and providing in home family therapy. After receiving her graduate degree she began working as a crisis counselor on a mobile mental health crisis team for Multnomah County. She began working at OHSU in 2008 as a resource social worker and joined the CF team in October of 2009. She is extremely excited to work with such a wonderful group of people. When Emily is not working she enjoys spending time reading, bicycling with her husband and playing at the park with her three dogs.

Patricia Rose, RD
Dietitian
Pat graduated from OSU in 1989 then attended her internship in Dallas, Texas. She has been working at OHSU as a pediatric dietitian since 1991 and has completed certification as a specialist in pediatrics. Pat is a National mentor for other dietitians in Cystic Fibrosis nutrition, an instructor in the School of Medicine, and trains dietetic interns in pediatric nutrition. Away from work, Pat loves boating, hiking, running, and being outdoors with her husband and two sons.

Wendy Palmrose, LC MSW
Social Worker
Wendy received a BFA from the University of California, Santa Barbara in 1978 and a counseling certification in 1980. She worked at Kerr Center for Handicapped Children as a child care aide, and then went on to earn her master's degree from Portland State in June 1987. After an internship in the Child Development and Rehabilitation Center, she was hired at OHSU and joined the CF team in 1988. She has been married to her husband Thomas, a respiratory therapist at Kaiser, for over 20 years, and they have a teenage son, Osten. Wendy is an award-winning in-line speed skater, an expert flower gardener, and secretly reads scientific articles.

Aaron Guzik, BA
Senior Research Coordinator
Aaron graduated from The University of Colorado in Boulder in 2005 with a degree in Integrative Physiology. While in Boulder, he began assisting in sleep medicine research, eventually moving to Boston, Massachusetts to work as a professional research assistant at the Brigham and Women's Hospital, where he continued working in sleep medicine, circadian physiology, and biological rhythms research. Aaron is a new member to the OHSU team and Oregon all together. When he is not on the hill he loves to cook, play ice hockey, run with his dog Bailey, and shop for kitchen gadgets.

Kim Keeling, RT
Lead Technician, Pediatric Pulmonary Lab
Kim runs the Pediatric Pulmonary Lab performing pulmonary functions, bronchoscopies, pH studies, and patient education. She is also assists with policy and procedure writings and implementations. She came to OHSU in 1993 after graduating from Mt. Hood Community College, primarily as an adult therapist, and was very involved in patient, staff, and physician education on respiratory equipment and therapies. In 2003, she began to train with Sara Rae in the pulmonary function lab, and upon Sara's retirement, took over lead tech duties. Kim loves working with our CF families, and also enjoys house remodeling, painting, and most of all, working with children. She is currently working on her bachelor's degree in business management.

3 The Emotional Aspects of CF

In this chapter:

- Dealing with a new diagnosis
- Telling family and friends
- Taking care of siblings
- Healthy ways to cope

"While raising our son with Cystic Fibrosis we've always felt it was imperative that he not only do all his required treatments, but that he know why each one is important. You never quite know as a parent if your child fully understands the lessons you are trying to teach. Then when Matthew was thirteen years old he was asked to do a short web video for a site called cfvoice.com. We were not allowed to be in the room while he was interviewed and his knowledge and feelings about the disease came directly from him. It was not until the interview went online that I realized just how much my son understands his disease and how it can affect him. Here are a few of the things my son shared in the video:

> *'I take my enzymes and extra vitamins when I eat. They keep me healthy. Eating will affect my lungs because nutritional health for CF people is lung health. Keep doing your treatments, keep taking your meds and stick with your dreams. Follow your dreams. Don't get down because you have CF. Just keep with it whatever you are doing.'*

I am very proud of our son and the way he deals with his disease. I am even more proud of him for sharing of himself to others who face the same obstacles."

Chapter 3
The Emotional Aspects of CF

The New Diagnosis

As a parent of a child who is diagnosed with CF, you may experience a flood of emotions. These emotions are normal and can range from anger, denial, shock, grief, helplessness, confusion, despair, sadness, and fear. All of these emotions are part of a grieving response. Some other emotions you may feel are worry, concern, guilt, and resentment. Emotions may come and go, reappear and change unpredictably. They can be triggered during unexpected times.

Upon hearing of the new diagnosis, parents might mourn the loss of the perfect, healthy child that they had hoped for. Sometimes they feel relief to know there is a reason for their baby's sickness. This sensation of relief may even make a parent feel guilty.

When a baby is diagnosed as a result of newborn screening, the opposite may be true. The baby seems wonderfully healthy. Being told about the CF diagnosis can come as a total shock.

A single parent may feel scared, helpless, and very much alone.

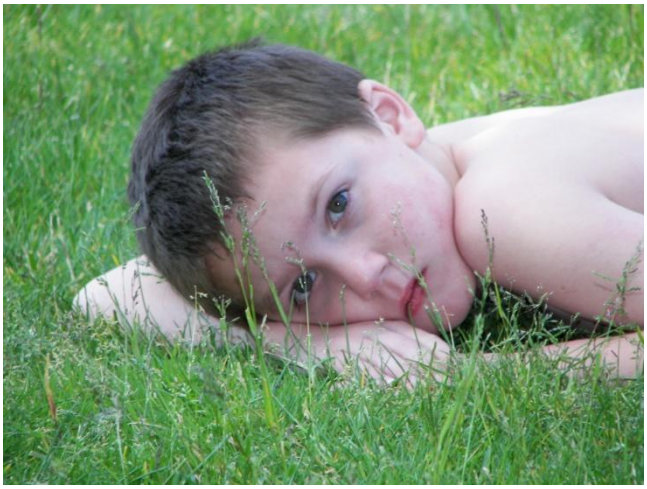
Relationships

In order to maintain your relationship, try to keep the lines of communication open. You are in this together. It may be difficult to talk about CF with each other because strong emotions tend to make it too hard to listen and give support. Each person could react very differently to the diagnosis and may experience varying reactions at different times. Discussions are even more challenging under these circumstances. This can put a strain on the relationship. It is hard to be supportive to each other when you are emotionally pained. Encourage each other to find additional and separate support systems. The more support you have, the better you'll be able to handle stress.

Family and Friends

Telling or 'breaking the news' to your family, relatives and friends about the new CF diagnosis can be difficult. A parent may initially only want a few family members to know, while another parent may feel comfortable telling everyone. Who you tell is entirely up to you and will depend upon the type of family you have.

Family, relatives and friends may not know what to do or what to say when they are told. No matter what their reaction is, it is important that each family member have someone else to confide in, where their feelings are safe to be shared. It is important to try to be nonjudgmental about each other's reactions.



Your family and friends who want to provide the greatest support to you can sometimes be a source of stress. Be sure to clearly communicate to them what they can do to be helpful to you. They may mean well, but if they don't know what to do, you may end up taking care of them rather than yourself.

Another person, outside of the family, may be able to help you get through this difficult time. Strongly consider finding a special confidante to talk to who will only listen, not try to offer solutions. Finding a support system that works to fit your needs is very crucial.

Eventually as you and your family learn more about CF and see the child doing better, the stress will lessen. It may take awhile to fully understand the disease of CF but increasing your knowledge and creating a strong support system are the best things you can do.

Siblings

Taking care of a child with CF can take a lot of time and attention away from other children in the home. The diagnosis of CF affects the whole family and it is normal for siblings to experience a wide range of emotions. However, children do not have the ability to fully understand their feelings.

At times CF will cause tension between siblings for a variety of reasons. The child with CF may resent siblings for not having it, while the child without CF may resent the extra attention the child with CF gets. They may be jealous of each other, even angry.

To help prevent such tension, consider involving your other children in the care of your child with CF. Siblings often want to help care for their sibling with CF. This also can allow them to begin understanding the disease. Many older siblings get protective and wish to monitor the health of their sibling. Be careful not to allow them to take on too much responsibility.

When the child with CF becomes sick, it affects the siblings. They can be worried, anxious or scared and may not be able to express their feelings. Try to talk openly about CF, including the medications and treatments.

Many siblings of children who have CF have said that they sometimes felt like they were invisible. Always try to consider spending special alone time with the child without CF. Remember that these siblings have unique personalities, special interests and needs that are separate from their brother or sister with CF.

Families often have more than one child with CF. It is important to understand that each child with CF will be affected differently. One sibling can be relatively well, while the other has more illness.

Daily Life

CF care requires a vigorous schedule of daily treatments and medications, and frequent clinic visits that disrupt normal routines. All this can impose significant stress on the child with CF, the parents and any siblings. Understanding the reactions to this stress and learning about effective methods to cope with them are important to the care of the child and to the function of the family.

As a whole, children with CF should be expected to live a very normal life. You should have the same expectations and give them similar responsibilities as other children. Try to avoid being overprotective so that you raise a happy healthy child.

Children with CF attend school, participate in sports and activities, play with other children, do household chores, and travel on family vacations. There are many aspects of daily life that are not changed by CF. It is very important for the child and family's emotional well-being that daily life is approached as normally as possible. One of the most valuable gifts you can give your child with CF (and yourself) is to treat them as a normal child who happens to have CF.

Educate

In addition to educating yourself, you will need to educate others about CF. The more people around your child that know about CF, the better, so be prepared to educate them. Some parents struggle with when, how and what to tell acquaintances about CF. They often worry that if others know the child has CF, their child will be treated differently.

However, it is important for those who come in contact with your child in the community (school, friends' homes, church, etc) to understand something about CF and share the same outlook of hope. By educating others, you can correct any misinformation on CF and develop a strong community support system for your child.

Most people do not know very much at all about CF so you may receive questions that are hard to answer. But each successful encounter can make the next one easier to handle. An honest, simple explanation is essential because it will set the tone for how others react.

At times when you are out in the public with your child who might need to cough, you might receive reactions from nosy and insensitive people. You or your child may get unwanted questions, very inappropriate comments and/or stupid responses, such as "does it affect their brain or intelligence?" "They don't look sick." "How long will they live?" "Is that coughing contagious?" "Don't they die from CF?"

When dealing with unwanted questions or comments, you can choose to ignore them, smile or shrug, briefly say that your child has a lung problem that causes coughing, or if you want, you can explain a little about CF. Always remember that just because your child has CF does not mean you are obligated to tell everyone.

Coping

People have different ways of coping with life's challenges. Some ways of coping can be healthy and some can be unhealthy. A common parental reaction, upon learning that your child has CF is denial—because it seems so overwhelming. Denial as a form of coping can serve a purpose, but only for very short periods of time. Denial acts as a protection in a stressful situation, but for longer periods it can be harmful and unhealthy.

When you have a child with CF, you are faced with accepting that your child has a chronic disease and you cannot change it. However, you can focus on how you can better cope with the stress that CF brings on.

You might consider thinking about developing positive ways to cope, (and how to deal with negative feelings), both now and into the future.

Here are some suggestions on healthy ways to cope that others have found helpful:

- Talk with someone you trust about your feelings
- Have an activity (walking, bicycling, exercising) or sport you enjoy
- Listen to your favorite music, watch an entertaining movie, read a good book
- Take time to connect with friends
- Find quiet places to rest or relax, think about your feelings, write in a journal
- Try to eat well-balanced meals and get regular sleep

Having a child with CF is very stressful; it may at times consume much of your thoughts. Use caution and try not to "bottle up" your feelings and worries because those emotions will be like a "time-bomb," surfacing when you least expect it, and maybe even when you have the least control.

You may find times when you have tried healthy ways to cope with stress but you just don't feel like yourself. You might need help in coping.

Signs of this may include:

- Constant thoughts or fears related to CF
- Inability to sleep
- Having moods swings with sadness, anger or anxiety
- Isolating yourself from family and friends, or having them worry about you

Counseling Resources

Sometimes life takes its toll on even the strongest person. It's common to seek out counseling to help alleviate stress. Counseling can be helpful to parents and other family members experiencing a wide range of challenges. These challenges may include: difficulty accepting the CF diagnosis, relationship problems, feelings of depression or anxiety, sibling struggles, financial stress, and overwhelming emotions.
It is important to recognize that depressive feelings can make it difficult to function in daily life and can interfere with the ability to work, sleep, and take care of yourself and your child. These feelings should not be ignored; there is an excellent chance that you can get effective help through counseling, or even with medications. If you are experiencing difficult emotions that make it hard to carry on with your life, know that you are certainly not alone; other parents have felt this way, too.
It is very acceptable to go to a counselor, many celebrities brag about seeing their therapists. Some people go to counseling just to have someone to talk to about life issues before they become major problems. Once you have entered counseling, it is usually viewed as a relief. The hardest part is deciding when and finding out where to go.

Contact your insurance company for information about mental health benefits. Some insurance plans will cover only licensed counselors and therapists with specific degrees, while other plans include a range of therapy professionals. A counselor, therapist or agency can also check your benefits and coverage.

Often, therapy professionals or agencies offer a sliding fee scale, meaning they charge based on your income and what you are able to pay. This includes those who have no insurance. There are several agencies that have low cost counseling.

There are different types of counselors and therapists who are licensed through the State. The important thing is to choose the right one for you. You can do this by interviewing them or asking questions about their practice, area of expertise and availability.

Some questions you may consider asking a professional or agency are:

- Do they have evening or weekend hours?
- Do they know about chronic illness? (They probably will not know about CF, but ask).
- Do they have special training in the area in which you need help?
- Do they work with individuals, couples, and/or families?

Sometimes antidepressants and other medications can help get you through a very difficult time. This is generally a decision made between you and your own doctor. If your counselor recommends them, then you may need to go to a psychiatrist who is the only professional, besides a doctor, licensed to prescribe medications.

4 Living with CF

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Chapter 4
Living with CF

4.1
INFANT TO 6 MONTHS

The arrival of a baby can be wonderful and exciting, but it can also be somewhat stressful and scary. Especially if your child has Cystic Fibrosis, which can present more challenges. Hopefully, through this guide and working with the CF Clinic, your transition to being a parent of a child with CF will be smooth and low-stress.

Step one - Enjoy your baby!
Sometimes a new diagnosis of CF can overshadow the wondrous joy of becoming a parent. The excitement and thrill of welcoming your new baby into the family should be the top priority. Your child is not a patient; your child is a person and you should enjoy all of the usual activities associated with having a newborn. And yes, that includes changing diapers!

The CF Clinic
The CF Clinic at OHSU is accredited by the National Cystic Fibrosis Foundation and treats patients from all over Oregon and SW Washington. One of the best things you can do to keep your baby healthy is to stay in close touch with the staff of the CF Clinic. The RN Coordinator will be a key person to establish a relationship with. The RN is available for advice and questions by phone and email.

Newly-diagnosed infants with CF are asked to come to the CF Clinic about every 1-3 months, or more often if necessary. You will also need to establish care with a pediatrician for non-CF care, such as immunizations and well-child checkups.

Growth and Development
The biggest health priority for infants with CF is growth and nutrition. Often, because of malabsorption, children with CF do not grow as well as other children. Your CF Clinic staff and pediatrician will be monitoring your baby's growth and making sure it's adequate.

One way your doctors monitor growth is by plotting your child's weight and height on a growth chart. This allows them to compare growth to other children and to track your child's progress over time. If you are concerned your child is not growing appropriately, ask your CF Clinic staff to show you the growth chart and discuss your child's progress.

At the CF Clinic, you will be seeing a dietitian regularly, who will take measurements and give advice on everything nutrition-related. This includes breast-feeding, bottle-feeding, formulas, vitamins, and medicines. When the time is right, you will be advised about the best solid foods to begin and how to maximize caloric intake.

Enzymes
Most children with CF need to take enzyme capsules to aid in the digestion of food. Although enzymes come in capsule form, you give them to your baby by breaking open the capsule and sprinkling the small granules in your baby's mouth with each feeding. The CF Clinic staff will teach you how. This medicine is crucial to your baby's growth and must be given with each feeding, whether it's from bottle or breast.

Vitamins
In CF, certain vitamins aren't absorbed well, so a multivitamin liquid will be prescribed. This is a special, prescription-only medicine and is designed specifically for kids with CF. It is high in vitamins A, D, E, K, and the mineral zinc, which can be deficient in children with CF.

Dehydration
One of the biggest dangers children with CF face is dehydration and/or low salt levels. Often, the CF dietitian will prescribe a little extra salt to add to your child's bottle.

Dehydration/low salt levels are not common, but it is important to recognize the danger signs. Please call the CF Clinic or your pediatrician immediately if you notice the following symptoms:

- Lethargy, sluggishness (less than normal activity)
- Irritability
- Sunken eyes
- Less urine in diapers
- Sunken fontanel (soft spot on top of head)
- Dryness of mouth
- No tears when crying

Malabsorption
Even with enzyme replacement therapy, sometimes a child with CF still does not absorb all of their food. When this happens, the child may exhibit some or all of these symptoms:

- Diarrhea, loose, or "pasty" stools
- Greasy stools – often indicated by an orange color or an oily sheen in the dirty diapers
- Gas, cramping – your baby will be fussy and stomach will be bloated
- Frequent stools – in excess of 4-5 per day
- Foul-smelling stools
- Bulky, large bowel movements

Constipation - An Important Consideration

Sometimes malabsorption can also be indicated by infrequent stools or constipation. This may seem confusing, but it has to do with the volume of stool produced in the intestine.

Important: If your child has not had a bowel movement for more than one day, it is vital you contact the CF clinic immediately. Severe constipation in a child with CF can be a medical emergency if not treated.

It's important that you do not adjust your child's enzyme dose on your own. If your child has a change in bowel habits, contact the CF Clinic for advice.

Lungs

CF also affects the lungs, which are prone to infection and inflammation. If your child develops a cough, has a stuffy nose, or sounds congested—or if your child is wheezing or has noisy breathing—contact the CF Clinic nurse for advice. Your child may be prescribed antibiotics to treat the lung infection

Traveling

Traveling with a baby who has CF isn't much different than with a child who doesn't. Remember to bring your enzymes with you at all times you never know when your child will want to feed.

If you plan on being outside on hot days, minimize the time in the heat as much as possible. Keep your child cool with loose, light clothing and stay in the shade. Hydrate with plenty of fluids—your dietitian can advise on you on the best selections.

Caregivers

It is important that anyone caring for your baby while you are away is prepared to deal with your child's Cystic Fibrosis. Of course, you will want to select someone who is trustworthy, responsible, and has experience caring for infants. Before you leave your child with the caregiver, it is helpful to have a training session to go over all of your child's medications and treatments. This should be done at a time when distractions are at a minimum; consider having a "practice" date with you and your child present so the caregiver can get comfortable with the regimen and ask questions. Have a list of phone numbers your caregiver can call, including the CF Clinic.

See the Nutrition Chapter for more information.

4.2
TODDLER

Managing Care for Toddlers
Having a toddler can be full of joys and challenges—having a toddler with CF is no exception! You have an amazing opportunity to establish a solid foundation of care for your child. What you teach and model now will have an impact on your son or daughter years down the road. Seize this opportunity and consider the following suggestions:

Routine
Every child has different needs related to CF but regardless of what your specific needs are, be sure to establish a clear routine. Help set expectations for your child so that he or she knows that the medications or therapies prescribed will be done every day.

Consistency
Life is busy and it can be challenging to maintain the routines you've started, but routines are only as effective as they are consistent. It does matter if a dose of medication is missed or a treatment forgotten. Consider ways to organize your family's outings or your responsibilities in such a way as to ensure the completion of the therapies that will benefit your child.

Parent/Guardian Directed

Right now all of your child's care is initiated by the caregiver. As your child continues to grow and mature they will become more and responsible for their CF. Developmentally, toddlers push boundaries and are exploring their limits. Your toddler may question an activity or try to take control by saying "no" to something you know will be best for them. As the parent/guardian, take a deep breath and set boundaries for your toddler. Find ways to offer to your toddler supervised control for meeting their CF related needs. Empower and involve them; for example, give your child the choice to choose between two snacks you approve of rather than asking your toddler what he or she would like to eat. Give your child the choice to do a treatment in five minutes or ten minutes. Have your toddler get supplies out or help you set up for a treatment. As the parent/guardian, the boundaries and expectations you set now will have a significant impact on how your child learns and manages self-care.

Advocate
Chances are that the people around you are not experts on CF. Some people may ask you uncomfortable questions or make uninformed statements. People usually want to be helpful but may have erroneous information or may be unsure of how to ask questions. Continue to educate yourself and in turn you'll be able to educate your family, friends and anyone else! You may be at a restaurant and giving your toddler high fat/high calorie foods which may be different than what someone else is feeding their child and that friend or restaurant patron may ask you a question.
Your answer is a great opportunity to advocate for your child and educate someone about CF. If your child stays with someone while you work, explain very clearly the needs your child has an advocate for

them in any situation. Feel free to ask guests to your home to wash their hands or use hand sanitizer. Assess the settings your child may be in and address any potential concerns that may impact your toddler's health.

Support

Equip family and friends with information about Cystic Fibrosis in order to help them understand what CF is and what it isn't. Pick a few people to train to care for your son or daughter, such as giving your child enzymes or respiratory therapy. The more people who can be involved with your child to offer support and encouragement the better! Learn to be comfortable giving medication or treatment with other people around so that you and your toddler aren't isolated—CF shouldn't be a secret or something to hide. It will be healthier for you and your toddler if you are able to involve others in this process. Likewise, begin to equip your toddler with language about CF. Explain to your toddler what you are doing as you prepare medications or treatment tools. These activities will be normal for your child so help them learn (slowly and appropriately) about their Cystic Fibrosis. These tools will help your child learn how to advocate too.

Affirm and Encourage

Your toddler will benefit greatly from positive reinforcement. Praise and encouragement you provide your child helps as you navigate CF together. Meeting your toddler's CF needs may be stressful at times, but the affirmation you give your child for cooperating when they take medications or participate in therapy will create a better experience for you and your toddler. Your child's CF needs will be specific to your toddler but when you affirm cooperation, interest, questions, and participation, you let your toddler know that you are proud of them and pleased with their much needed involvement.

A few other thoughts

As you navigate CF with your toddler, be sure to utilize the resources offered by the CF Clinic!

Whenever you have questions, contact the clinic. You are still learning about CF and your toddler; no one expects you to have all the answers. You aren't "bothering" the staff at the clinic by asking questions. You are being the best parent/guardian you can be for your son or daughter when you get the information to make informed decisions about your child's health.

Encourage exercise—make it fun! A trip to the park, playing chase in your home, running around outside, even modeling how you exercise are great examples for your toddler to learn. Exercise is great for kids with CF!

Experiment with foods and ways of eating—nutrition and getting enough calories is important so consider ways to make eating an enjoyable time.

Expose your children to new things and experiences! Your toddler, like any toddler, is curious and wants to learn. Meet that curiosity head on—introduce your toddler to lots of books, go on outings, talk to them about what is going on around them…toddlers are like sponges waiting to soak in everything you share with them!

4.3
EARLY CHILDHOOD

"One day our 4 year old daughter, Ella, was finishing up her chest physical therapy with her vest system when a friend arrived early for a play date. The friend climbed onto the couch beside Ella and just stared at her for a few moments. Then she gently put her hand over Ella's vibrating torso and said, 'Wow! That's shaky. Does it hurt?' My daughter smiled and replied, 'Nope. It just shakes my eyes too much sometimes.' This friend quietly sat beside my daughter, patting her vest every once in a while. As the PT session ended, Ella's friend said in all 4 year old sincerity, 'I have to get my mom to take me to the mall. I gotta get one of those vests too!' Leave it to the innocence of a child to make something so loud, jarring and out of the ordinary, like Ella's vest machine, seem downright cool."

Early Childhood — Managing Care at Home
Here you are with a child that sometimes acts like a toddler and sometimes seems so much more grown up than his/her actual age. Early childhood certainly comes with its challenges. Your child may be anywhere from three to six years of age, and in pre-kindergarten to second grade.

Here are some suggestions for taking care of your child at home:

Stay Positive
You are the model for your child. When you talk with your child about his or her disease, stay positive. Your child will follow your lead—a child learns to deal with their feelings by imitating others. Focus on your child's good behavior choices.

Include Child in the Routine
By asking your child questions like "What treatment do we do next?" and "How many enzyme capsules is needed for that snack?" you are including your child in his/her routine. Your child will gain confidence and you set stage for self-care.

Meds and Treatment —Explain the What and Why
Engage in conversation with your child about the medicines and treatments he or she is taking. For example, "This medicine is called Albuterol. It is to help your lungs." By explaining "the what and why," you help the child feel more in control and their CF care becomes less scary. When your child is able to talk about CF care they begin to accept it as a routine part of life.

Focus on infection control
Teach and reinforce good hygiene. Washing hands with soap and water and/or using hand sanitizer can steer you and your child away from a lot of illnesses. Make it a routine to wash hands before you eat, drink, or come in from the outside. When using soap and water, it is recommended to scrub with soap for at least 20 seconds.

Taking steps to control infection also means preventing it. The goal is to keep your child healthier and feeling better. It all comes down to following your child's treatment plan. Stick to the plan. If something seems wrong, talk to your CF Clinic team.

Reward System Examples
Children like knowing they are doing well and making smart choices. Reward your child when they complete their treatments and medications. There are many ways to do this. For instance, your child can choose which activity to do during treatment. Sticker charts are a great idea to enhance daily treatment compliance and cooperation.

Consider reading to your child during nebulizer treatments. This can make the sessions more comforting. Give choices such as "Would you like to start your treatment before dinner or after dinner." Question that are less effective are: "Are you ready for your treatment?" These questions set you up for the answer "no and a negative outcome. Instead, try: "Which book should I read during your treatment?" or "Which cartoon do you want to watch during your treatment?"

Give possible reinforcement when your child is compliant with treatments. Promote healthy choices. Talk with your child about what others do to stay healthy. For example, "Dad takes his blood pressure medicine to stay healthy," or "Mom does her exercises so she can be strong."

Other things to consider
Before going to see the doctor, consider helping your child think of one or two questions he or she wants answered. Avoid asking or answering for your child—let the doctor speak directly to your child. This sets the stage for your child to openly talk with CF team members.

Early Childhood — Managing Care Away from Home
Taking care of the CF child at home is tough at times. Then there are times when your child maybe at school, camp, or a sleepover. Here are some thoughts for managing the child's care when he or she is away from home:

Organization is Key
Make a schedule for your child's care. Write it all down—the meds and the treatments. Help your child have a consistent, routine treatment schedule that has some built-in flexibility. If your child is away from home without a parent/guardian, you will need to train one or two adults that are in charge. Keep in mind not everyone will be up to the task and they may not always be successful. Be patient and remember your child can start helping with their care. He or she should be able to answer some questions regarding their routine.

Make an emergency contact list. Include the names and telephone numbers of your child's doctor, a list of medications, therapies, and dosages and frequencies. Keep this list handy and remember to update it whenever there are changes

Travel Kit
Consider putting together a travel kit including the schedule, medications, and other materials your child needs for their CF care. This will help you feel more spontaneous and less likely to forget something necessary.

Adjust the Schedule
Because we want our kids to be kids first and not think about having CF all the time, make sure your child's routine can be flexible. Skipping treatments is not advisable; however, bumping up the schedule to accommodate a social activity is just plain smart. For instance, let's say your child has been invited to a birthday party at 4:00 pm, yet he normally does treatment at 4:30 pm. Don't skip the treatment; just do it before you leave for the party.

Focus on "Like Everyone Else"
Around age three to six, your child may start to notice that other kids do not have the same routine of medicines and treatments. Or they may cough more. Whenever possible, try to talk with your child about how they are like other kids. All kids like to run, play, draw, be loud, and have a good time. You can acknowledge your child's similarities with their peers by saying things like: "Wow! You and your friends are really good runners!"

You may also want to point out that other people have medical issues too so your child can become more comfortable with CF. For example, you can talk about how Grandma has a bad knee, a neighbor's child takes insulin, or Mommy needs special cream for her skin. Teach your child that we all have things we have to deal with in our lives. Help your child realize that we all have to cope with one thing or another.

4.4
MIDDLE CHILDHOOD

Middle Childhood—Managing Care at Home
Now you have child in grade school, approximately seven to ten years old, and on top of that he or she has CF. Your child still needs your care and guidance. Together you will continue to grow your understanding and coping skills with this disease. Here are some suggestions for taking care of your grade-schooler at home:

Stay Positive
Continue to be the model for your child. Keep a positive attitude even when your child is feeling down about their illness. Continue to acknowledge the good choices your child is making toward taking care of him or herself.

Everyone has his or her own way of coping. To reduce stress, be flexible. Obtain information and don't make a problem worse that it has to be. Sometimes just changing the way you look at a situation can make a huge difference.

Include Your Child in the Routine
Keep involving your child in the routine. Ask your child questions that help prepare them for when you are not with them. For example, your child should know how many enzymes they take for meals and how often he or she does CPT. This is also a good time for your child to be learning his or her doctor's name and hospital name.

Your child can play an important role in the timing of treatments. Work with your child to make a daily schedule for their CF care that fits around other important activities. This shows that CF care is important but so are other things in life. Keep the schedule flexible as possible because conflicts come up from time to time.

Increase Your Child's Level of Responsibility
Step up the level of responsibility your child has regarding his/her care. For example, your child can help clean their breathing equipment. Children want to know what is expected. Give your child a time when CPT has to be completed. This shows flexibility and puts the child in charge of when it gets done before a given deadline.

When visiting the nurse or doctor, encourage your child to speak directly to him or her. Let your child ask questions and talk about how he or she is feeling. Building a good report now will pay off later.

When appropriate, put your child in charge of carrying their enzymes for the next snack time. Allowing your child to be responsible from this way helps your child feel more like they are in control. It's also a great way to learn the consequences of their behavior.

Promote Self-Starting
Ultimately we want of children to learn to care for themselves. In middle childhood it may feel like that is a long ways off, but you can take steps now to prepare them. Keep asking questions like, "What do we do next?" and, "How many capsules do you think you need?" Allow your child to make choices and even mistakes. This is how we learn. Your child can begin to accept the consequences of their choices. For instance, if your child forgets to take enzymes before munching on three cookies, he or she may have a

stomach ache and/or loose stools. This experience emphasizes the importance of making a better choice next time, like taking enzymes before snacking.

Try New Reward Systems
When it comes to rewards, what worked for your toddler is probably not going to cut it for you grade-schooler. Ask other parents what works for their families. Basically you want to encourage the positive choices your child is making. Letting your child choose when he or she will do treatments can be a bonus.

Encourage More Awareness of the Body
Your child can begin to understand more and more about lung function. Talk to your child about how the lungs work and how CF problems can arise. This helps your child understand why the medicine and CPT are important. Your child can also begin to describe what he or she is feeling when starting to get sick. This means your child is on the way to becoming an expert on changes in his or her body that might indicate infection or other problems. Encourage your child to point out changes such as increased coughing, less appetite, less energy, or more mucus. Early detection of a problem can lead to earlier, more proactive treatment.

Expect More Talks about CF
In middle childhood, your child can begin to use correct vocabulary to talk about their care. Your child will grow to learn the names of treatments, equipment, and medications. Help your child to know how to handle questions and comments about CF from others. You may want to practice and role-play situations aloud so that your child feels more confident and comfortable talking about their disease.

Middle Childhood—Managing Care Away from Home
Your child during the grade school years is becoming more and more socially active. There may be birthday parties, sleepovers, day & overnight camps, sport activities in addition to school that keeps your child away from home. You may even find yourself not the main caregiver for your child for multiple days.

Here are some suggestions for managing care of your grade-schooler when he or she is away from home:

Make a List
Think about your child's daily routine. Write a list of pills, inhaled solutions, medications, and therapies. Record the times and dosage amounts to be administered. Include important phone numbers like yours, your child's doctor and the CF clinic health care providers. Go over this list with your child so they feel involved. Use this list to educate adults in charge when you are not with your child.

See the Resource Chapter for an example.

Train the Caregiver
Depending on how long your child is away from home, you will want to go over what your child is going to need. First pick an adult you can trust. Choose a time to train the caregiver when distractions are at a minimum. Consider even having a "practice" date with your child present. Keep in mind that it might be a little overwhelming at first for the trainee. Provide assurances and phone numbers. Remember as your child gets older they will be able to answer some questions asked by the adult in charge.

Consider Self-medication Experiences
When and where appropriate, consider letting your child carry a small amount of medication so it is handy. Start with one dosage. Although most grade schools do not allow students to carry their own medication, consider it for non-school events like birthday parties. If your child is carrying medication always let the caregiver in charge know. By giving your child the opportunity to medicate themselves helps them on the way to becoming more self-reliant.

Encourage Your Child to Advocate
Your child needs to be able to talk about CF. Encourage them to express feelings about treatments and illness. By talking at home, you are helping your child to be able to speak about his or her disease anywhere. Your child can learn to handle comments and questions about CF. Explore with your child using role plays to discover ways to talk with people. If your child can express feelings and provide information they will feel more comfortable and confident.

Encourage your child to talk to his or her teachers. Communicating with the teacher directly rather than through parents all the time is an important step toward independence. For instance, your child at certain times needs to be able to tell an adult other than a parent what is going on with his or her body. Things like coughing, tummy aches, gas, and diarrhea can be embarrassing. You want your child to be able to get help when they need it. CF children need to be able to recognize a problem, ask for help, and feel better quickly.

Stand Back and Let Your Child Ask & Learn
As your child becomes more involved with their CF care, they will need more information. By obtaining more information, you child begins to understand the "why" behind some things. This makes it easier for your child to accept new responsibilities. At clinic visits, encourage your child to ask questions, share information, and figure out what else they want to know. Stand back and let your child talk with members of the CF team. Support your child in becoming an expert when it comes to his or her own body.

4.5
TEENS

"We have two daughters with Cystic Fibrosis, Hayli, age 18, and Kendra, age 16. Our message is one of encouragement. Support your child with CF to live life to the fullest. For some that may mean pursuing a passion in sports or playing an instrument. Whatever you do, don't let fear of the unknown hold you back from letting your child experience and participate in life activities!

Among being very active teenagers and participating in many different activities, Hayli has played the clarinet for eight years, and Kendra has played the trumpet for six years. When Hayli was in the fifth grade, and she wanted to play the clarinet, we encouraged her to give it a try! Kendra followed in her footsteps, but was interested in the trumpet and we supported her as well. Hayli has taken her music to great heights and has participated in marching band and symphony for the last four years, even taking a trip with her wind ensemble group to Indiana to play in a special concert. Kendra played in her middle school jazz band and now plays in the high school symphonic band. It seems as if band might be an odd choice for someone with CF, but we did not hold the girls back, and it has brought them and us great joy.

Kendra participates in track and cross country. She loves to run! Exercise is very important, and Hayli works out at the gym as much as possible. They have many friends, have gone to the annual snoball dance, senior prom, and had braces and wisdom teeth pulled. Hayli has a driver's license and Kendra is learning to drive. Both girls have busy, active lives, and they don't let CF hold them back! CF is part of their lives, but it doesn't define who they are, or what they dream of becoming."

Teens - Managing at home

The teen years are a time of great transitions for both children and their parents. For teens it is a time of intellectual and emotional flowering. It is a time of testing boundaries, searching for new experiences and increasing independence and responsibility for their own care and actions. For the parent it is a time of letting go, and trusting their child to make appropriate choices. The teen years also present times of worry and conflict. Keep in mind; this is a normal and necessary stage of life, which we all pass through on our way to adulthood.

- **Transitioning from Dependence to Independence**
  The family with a child with CF, confronts all the normal opportunities and challenges of teen life, plus the day to day challenge of managing CF care. As with other aspects of life, the teenager with CF should be increasingly responsible for their own care and behavior. However, your teen is not yet fully an adult and the ability to put off immediate gratification, in the interest of long term well-being, may still be somewhere on the horizon. It is as important to allow your teen to take responsibility as it is to make sure the responsibility is carried out. With respect to CF care, this can be as simple as, for example, allowing your teen to initiate their care and treatments, but

confirming that the therapies have been done. Remember, this is a learning process and won't happen perfectly from day one.

- **Maintaining Care in a Busy Schedule**
  Scheduling respiratory therapy, medications and other treatments in the busy life of a teenager may present difficulties. Late nights, after school activities and spending time with friends may all combine to make time for treatments a seemingly random and infrequent occurrence. Flexibility is the order of the day. It is more important that treatments occur, than *when* and *where* they occur. Work around your teen's activities to find times when treatments can occur. Encourage treatments to be a part of group activities rather than something that is done alone. Peer support can be a major asset in insuring that your teen's therapies are perceived as normal and not stigmatized. It is imperative for you teen's long term health that effective care and treatment continues, even while so many other things seem to take precedence.

Teens - Nutrition

With the onset of puberty the nutritional needs of children with CF increase dramatically. Teens with CF may need 30—50% more calories than other teens. Eating 3,000—5,000 calories daily can be a daunting task, but it is important! Good nutrition fuels the body's change from a child to an adult. Good nutrition also has a positive effect on lung function. A high Body Mass Index (BMI) is associated with better lung health.

For teens with CF good nutrition means:

Eat a balanced, high-calorie diet with plenty of fat and protein.

High-Energy Lunches:

* Double cheeseburger, french fries, apple, ice cream
* Fried chicken, potatoes with gravy, vegetables
* Pizza with extra cheese, orange
* Macaroni with extra cheese, grapes
* Double ham-and-cheese omelet with vegetables
* Deli-sandwich with extra cheeses and meats

High-Energy Snacks:

* Mixture of nuts, raisins, dried apricots
* Apple pie with ice cream or cheese
* Strawberry shortcake with whipped cream
* Banana split
* Cheese, crackers, nuts
* Peanut butter crackers, muffins, bagels
* Frozen yogurt or ice cream with nuts and/or candy pieces
* High-calorie sports snack bars

High-Calorie Milkshakes:

* Homemade milkshakes with ice cream, cream, powdered milk, or instant breakfast powder, with fruit, or store-bought, calorie-rich shakes

Pancreatic enzymes help digest and absorb food. Take enzymes just before eating a meal or snack. Not taking enzymes may lead to trouble gaining weight and height, more frequent, foul-smelling bowel movements and an intestinal blockage, called distal intestinal obstruction syndrome.

Multivitamins that have more easily absorbed forms of vitamins A, D, E, and K are generally prescribed for people with CF. It is crucial that the vitamins prescribed be taken daily. Take vitamins with enzymes and food to aid absorption.

Teens - Low Appetite

Many people with CF struggle with a low appetite and don't feel like eating. When you need to eat 3000-5000 calories a day this can be a problem! Because they have problems digesting fats and proteins they may prefer lower calorie, low fat foods. Violent coughing associated with exacerbations and antibiotics may leave them experiencing nausea. Gastroesophageal reflux (heartburn) and gas may make eating uncomfortable. In addition to taking enzymes with every meal the following may help to increase caloric intake:

Eat regular meals together: Eating is more enjoyable in a social setting. Eating together can allow a parent to make sure their teen is getting good nutritious meals and taking enzymes and medications on a regular basis.

Provide high calorie snacks: Have high calorie snacks available at home and when your teen is on the move. A fast food restaurant is an excellent source of high fat, high calorie food!

Exacerbations and Antibiotics: Keep weight in the 50th percentile or above when feeling well to counteract weight loss during exacerbations. Take probiotics such as acidophilus to restore intestinal bacteria.

Gastroesophageal reflux: The CF Center's Health Care Team can prescribe acid reducers.

Liquid high-calorie supplements can be added to diets to increase caloric intake. The CF Center's Dietician can provide recommendations.

Feeding Tubes: Many people with CF cannot eat enough food to meet their daily caloric needs. An additional 1000-2000 calories per day of high calorie liquid supplement can be fed through a tube directly into the stomach. Many teens with CF get calories through a feeding tube with a pump all night in private, and then have normal daytime meals and activities. Talk with the CF Center's Health Care Team to learn more about tube feedings**.**

CF-Related Diabetes:
Some teens and adults with CF also have CF-related diabetes. Lung infections and steroids can make blood sugars go up. If blood sugar levels stay too high, a diabetes specialist will help to decide how to control blood sugar.

Exercise
Exercise strengthens lungs, helps to clear lung mucus, builds muscles and strong bones, increases appetite and can impart a feeling of well being. Brisk walking, running, swimming, biking, and jump roping are aerobic exercises that can benefit teens with CF. Exercise about 30 minutes, three times a week and don't exercise too hard. People with CF lose more salt while sweating than people without CF and should eat high-salt foods like pretzels, chips or salted nuts and drink plenty of water, fruit juice, or sports drinks before, during, and after exercise

Tobacco, Alcohol and Drug Abuse
Most teenagers are likely to engage, at some time, in the use of alcohol, tobacco or recreational drugs. This is as true for the teen with CF as any other teenager. The dangers of alcohol, psychoactive drugs and smoking are widely known, but teenagers still engage in these risky behaviors. Indeed, because of the developing nature of the teenage mind, new risk taking experiences are particularly attractive to them. However, an understanding of the potential consequences may be elusive. What the teen with CF and their family should know is, the consequences of tobacco, alcohol or drug use can be particularly devastating for someone with CF. The dangers of smoking can readily be understood by most of those with impaired lungs, the danger of alcohol and other drugs may not be so apparent. The liver and other organs of a person with CF may also be impaired. In both cases permanent organ damage may result at a level of use generally tolerated by non-CF persons. The CF Center's Health Care Team can assist in finding resources for patients that need treatment for tobacco, drug or alcohol abuse.

Sexuality
Intimate and sexual relationships are likely to begin during the teen years. Teens with CF are not an exception. Pregnancy and Sexually Transmitted Diseases (STD) are as much of (or more) of a concern for the teen with CF as those without.

Females with CF are fertile, although some may have difficulty conceiving. Before becoming sexually active, women should talk with their health care provider about birth control and what methods will work for them. Many antibiotics interfere with the effectiveness of birth control pills, therefore additional protection is needed. Safe sex practices including condoms are advisable.

Males with CF (most, but not all) are generally considered infertile. This does not affect their ability to have sex. Men with CF produce sperm in the testes, but delivery of the sperm is blocked in the vas deferens. It is possible for men with CF to be biological fathers. Current medical technology can harvest the sperm in the testes, which can then be used for artificial insemination. For sexually active males safe sex practices, including condom use, are advisable.

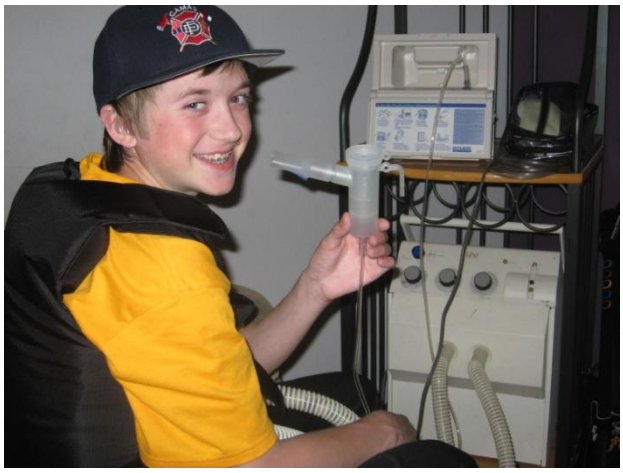
Pregnancy for the women with CF should be a well-planned happy occasion not an unwanted surprise. Nutritional status, lung exacerbations and general health can all affect the well being of the mother and child. Women with CF who are planning to, or have become, pregnant should talk with their CF Center's Health Care Team. All partners of men and women with CF, who are planning on having children, should be tested to see if they are CF carriers.

Teens - Away From Home

The opportunities for travel, camps, and other extended activities away from home and parents become more common in middle and high school. These opportunities can be significant, life changing experiences for your child. Having CF should not be a reason to prevent them from participating in these activities. Careful preparation will allow your child to fully participate without undue risk to their health. Just like at home, it is important that medications are taken, R.T done, and adequate food intake and rest occur. This can be especially difficult while traveling. Here are some things that may help make the trip go more smoothly:

Make sure to start out healthy! Schedule a clinic visit, a few weeks before the event, to allow time for treatment, if required, before the departure date. **Familiarize the responsible adults with the care regime**, medications and other special needs of your teen. Review emergency procedures and contact information. Write all of this down! Include your teen, they should be aware of all arrangements.

Review the itinerary or schedule and create a treatment schedule that fits within the general program. Find times when therapies can be done without taking time away from other activities.



Be flexible, it is important that your child can fully participate in all of the activities as well as find time for RT and taking medications. It is more important that therapies are done than they are always done on time. The schedule is likely to change and your teen and responsible adult will need to "fit things in" on an ad-hoc basis.

Organize medications in a safe and convenient way to distribute during the day. Keep all medications and equipment, as possible, in a carry-on bag if traveling on an airplane. Keep a list of medications, prescription information and your CF Care Center contact information available at all times. Have

extra medication and equipment available to replace those that become lost or broken.

Prepare for exacerbations or other emergencies: If an exacerbation occurs while traveling have the appropriate antibiotics on hand for immediate treatment, if possible. Identify CF care centers along the way and locate pharmacies where you can get additional medication. Make sure that there are enough adults to allow, at least, one to accompany your teen to the hospital if necessary.

4.6 ON THE WAY TO ADULTHOOD…

Walking to School with 65 Roses
by Hayli, Age 19

"Cystic Fibrosis, a disease not many have heard of, and yet a disease that I and many others live with everyday. One aspect that has always been difficult for me has been trying to explain the disease to people who see me taking my pills and ask, "What are those for?" It's not that I don't want to explain why I take enzymes, but although I have CF I have never fully understood exactly what causes it, other than that both of my parents happen to carry the CF gene. So here's my typical response for when people ask: I have Cystic Fibrosis, or CF (this is usually followed by a confused expression from my peers, which sometimes makes me laugh), and then I explain to them that it's a genetic disease which affects my lungs, makes me cough more, and I need to take pills before I eat in order to help digest my food because my pancreas doesn't work right. Yep, in a nut shell, that's basically exactly what I tell people when they ask. It's not too scientific, but after that brief explanation people seem to understand.

School for the most part was never too difficult in the terms of dealing with CF. Yes; I had my sick days where I would miss up to a week of school at a time. Fortunately I was blessed with wonderful, caring friends who would pick up homework assignments for me and help me catch up. My friends are what made school not so difficult—just having one close friend to confide in makes all the difference in the world. Not all of my friends knew that I had CF, but the ones that I felt truly cared, I told, and because I told them my life became that much easier. I had people that I could talk to (some more than others, I didn't tell all of my close friends about every aspect of my CF). Whenever there was a CF walk sponsored by the Cystic Fibrosis Foundation, I often had as many as five friends join me.

I had to go to the hospital a total of three times during high school, once during my junior year and twice my senior year —I got my PICC line removed the day before my high school graduation. Hospital stays, naturally, were never something that I looked forward to but as I got older, especially in the last two years of high school, I realized that they were for the best I knew that in order to be the best version of me that I could be, hospitalization was necessary and once again, friends made those stays better. Each time I was hospitalized I stayed there for about two or three days and then went home for IV therapy. Going to school wasn't too bad with a PICC line. My family and I let the school nurse know, and I usually let one or two friends know as well. I wore sweaters to cover the PICC line, which worked amazingly well. The nurses also worked up a treatment schedule so that I only needed to be hooked up to the medicine right before school, right after school, and later at night so I never had to miss any class because of it or do any of the treatment during school hours which was quite nice to say the least.

Other than just friends in high school, I did have a couple of boyfriends (not all at once of course). The most important thing that I learned from dating is to tell the person who caught my eye and made my heart race, that I had CF right off the bat and I also told them that if they had any questions they should ask me, because the internet can be full of lies and is often outdated. Luckily enough the guys that I happened to fall for were really great when I told them However if I had ever met a guy that, for some reason or other, could not

handle the fact that I have CF, I suppose that I would be glad that I told him sooner rather than later so that I could realize that he's not the guy for me, and kick him to the curb.

One of the hardest parts for me health wise during school was towards the end of my junior and senior year I started coughing up blood. , I didn't cough up significantly large amounts at a time, but it would happen quite frequently which was frustrating to say the least. I participated in marching band all four years of high school and when I started to cough up blood, in I needed to take a break from physical activity to heal the hemorrhaging vessels. Dr. Powers always described it as a double edged sword; on one hand I need to exercise and do my treatments to stay healthy, and on the other hand if I do I will cough up blood which is not a good deal. Needless to say I have taken a lot of Zyvox, which usually helps. Last September I had the embolization procedure I had been wanting for a very long time. The procedure was smooth and easy ;it blocked the blood vessels in my lungs that were enlarged and causing the bleeding when I coughed and I hoped that I would stop coughing up blood at least for while.

The procedure could not have come at a better time;, I was starting (Western Oregon University) and one of my fall term classes was dance aerobics—a class I wanted to fully participate in. Before I moved in with my two roommates, we talked online and I let them both know about my CF and the therapies that I have to do, mainly Pulmozyme, the pills that I have to take, as well as the fact that I'm a cougher. Both of them were extremely welcoming and friendly and the fact that I have CF did not seem to bother them in the least. So far college has been an amazing experience and I would not trade it for the world.

Some people say that they are proud of me for "going through this." What I tell them is that this is my life, things are how they are, and I can't imagine living any other way. To others, CF is a separate entity hindering my life; however for me, CF is a part of me, like an ugly birth mark. I may not want it, but it will always be there, and so why let it bother me? I'm not going to let CF control my life or cause me to worry about what may be, or what will happen to me in the future. I simply live everyday to its fullest, and take a large gulp of air as I plunge into the next."

5 Respiratory Care

In this chapter:

- Airway clearance techniques
- Medications for better lung health
- Pulmonary function testing (PFTs)

"When my husband and I were children, people with CF had very little hope of living beyond their teenage years. Today, with earlier diagnosis, more effective therapies and drug development, people with CF live well into their middle age and older. We have a son with CF and we hope he has a long and fruitful life. There seems to be many breakthroughs on the horizon that would help our desire come true. We have many other things to be grateful for as well. Evan is not only dearly loved by his family, but he has love, kindness and support from his many friends and their families.

Evan is now 14. When asked what helps him get through his days, what has been the greatest source of hope and support, his immediate response is, 'My friends. They treat me like I'm a regular kid. My CF doesn't matter.'

Evan's response is typical of any young teen. Friends are the most important things in their lives. We are very grateful to his friends for understanding and helping him. Evan keeps stashes of his pancreatic enzymes at all of his friends' houses. He has frequent sleepovers, excursions and camping trips with his pals. He is managing his disease, and managing to make and maintain friendships that are true and long-lived.

He has also been very open and honest about his disease. From a very young age he has tried to understand the complexities of CF. He has also been active in informing people about the disease, giving his first in service to his teacher and classmates, in first grade. Evan knows that he has to sometimes advocate for himself. This is a great skill for a child to have.

Evan's medical caregivers have also worked at understanding that he is a child first. They have always included him in their conversation, helped him voice his opinions and choices in therapies and care. They have made sure they didn't ask too much of him, so that his childhood and development could play out well balanced.

With this support we hope to see Evan go on dates, go to his Senior Prom, graduate from high school, go away to college, travel the globe, become the professional he chooses to be, and perhaps get married and have children of his own. The friendships and relationships that he has now will help him do all of these things and more.

Help your child be a child. Help your kid with CF understand that he or she doesn't stand alone, that there are many people in the world that are willing to help them be themselves, the best kid that they can be. Cystic Fibrosis doesn't have to get in the way of that, in fact it may be a way for your kid to grow up in many ways that many children do not."

Chapter 5
Respiratory Care

Cystic Fibrosis is characterized by the production of thick, sticky mucus that clogs the airways—leading to chronic infection and inflammation. Over time, it leads to lung damage. Fortunately, there are a number of treatments and medications you can do to lessen the damage and keep your child as healthy as possible.

AIRWAY CLEARANCE (CPT)

One of the key ways to minimize lung damage is to try and clear the sticky mucus from the airways. This is called ***airway clearance,*** or sometimes ***chest physiotherapy*** or just ***CPT***. This can be accomplished in many ways, and not all treatments are appropriate for all age groups. Your CF team will give specific instructions on the best technique for your child, and when it's best to start. Examples of different methods are outlined below.

Clapping

This technique is appropriate for all ages, including infants, but *should only be performed after careful instruction from your CF team's respiratory therapist.* Gentle pats are given in a rhythmic fashion to the chest wall, sometimes with soft, suction cup-like paddles. The idea behind this and other CPT techniques is to loosen the thick mucus from the walls of the lung tubes, allowing your child to cough it out and clear the airways.

Positive-Pressure Devices

For older children who can follow directions, this technique is very effective for airway clearance. It involves blowing into a device that offers resistance. The resistance creates a backwards flow of air into the lungs which loosens the mucus. There are a few different manufacturers of positive-pressure devices, which have names such as PEP, Acapella, and Flutter. A big advantage of this method is that a child can perform this maneuver on their own, without a second person.

Percussion

Another method of loosening mucus is percussion with an electric percussor. Similar to a vibrating massager, the percussor vibrates the chest wall and shakes the mucus loose. This requires another person, like a parent, to administer the treatment.

Therapy Vest

A therapy vest uses compressed air to deliver the vibrating, loosening movements via a vest or jacket worn around the chest. Its advantage is that it's hands-free and can be used on small children as well as adults. However, it's an expensive device, and many insurance companies don't cover it.

Huff cough

A huff cough is not a separate therapy—it is used in conjunction with any of the therapies above. It's a specialized coughing technique designed to effectively clear the loosened secretions. When the time is appropriate, your CF Clinic respiratory therapist will teach your child proper huff cough technique.

Parents often ask what the best CPT method is, but there is no "best." Studies have shown that all of the techniques work equally well, but the key is that they need to be done consistently and regularly. CPT

should be done daily, and often is best if done two to three times a day. CPT is one of the simplest, cheapest, and most effective way to keep your child with CF healthy.

CPT is not appropriate for all patients with CF, and not all methods are appropriate for all patients. Please ask your CF team which CPT method is right for you and if your child should begin treatments.

MEDICATIONS

Respiratory care also involves giving medications—many of which have been designed specifically for patients with CF. Your team will prescribe medications based on your child's individualized needs—not all medications are appropriate for all persons with CF. Ask your CF team physician or RN if you have questions regarding medication. Some common ones are outlined below:

Pulmozyme

A mucus-thinning medication. It's a liquid medicine put in a special machine that turns it from a liquid to an **aerosol,** or mist, that your child can breathe into the lungs (**"nebulizer"**). Pulmozyme is given daily as a long-term, maintenance therapy. It is usually not prescribed until your physician sees lung symptoms from CF.

TOBI

An antibiotic. TOBI is also an inhaled medicine that is given by nebulizer. It is **only** for people who grow a specific bacterium in their sputum (*Pseudomonas aeruginosa*). TOBI is given twice daily as a long-term, maintenance therapy.

Hypertonic Saline

A mucus-thinning medication. Also helps to hydrate the thick mucus. Also inhaled with a nebulizer, it's given once or twice daily as a long-term, maintenance therapy.

Azithromycin

An antibiotic. Azithromycin is given by mouth, prescribed to be taken three times a week for CF patients. It fights against the growth of bacteria in the lungs for patients who grow *Pseudomonas aeruginosa.*

Ibuprofen

An anti-inflammatory. Ibuprofen is given twice daily in therapeutic doses. It fights against the inflammation occurring in the lungs.

> ***WARNING:*** *Ibuprofen is available over-the-counter, but can be VERY DANGEROUS if given in a way other than what is recommended on the product label. Your CF physician will order special testing to see if your child qualifies for this therapy.* ***DO NOT GIVE this medicine until consulting with your CF team.***

Inhaled Steroids

An anti-inflammatory. Only for certain patients who have a reactive airway component to their CF. Generally given twice daily.

Bronchodilators
Open up the airways in certain patients who have a reactive airway component to their CF. Given as needed, often before other treatments such as Pulmozyme or TOBI.

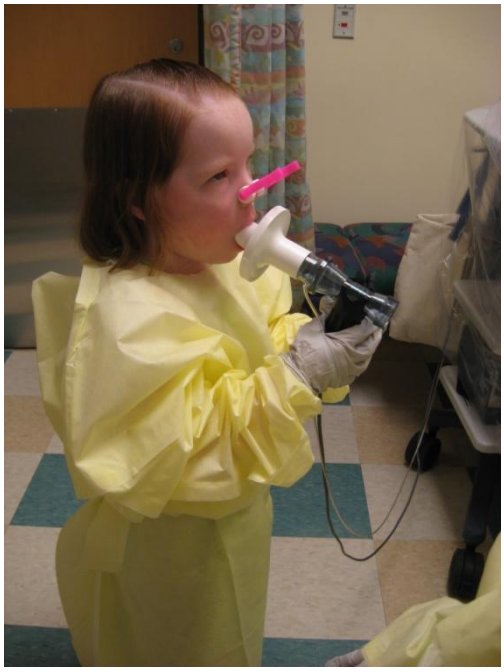
The order of inhaled medications and treatments is important. If your child is on more than one medication, it is important to give them in the correct order to maximize benefit and prevent complications. Below is an example of the typical order of all of the above respiratory care treatments:

- Bronchodilator
- Pulmozyme or Hypertonic Saline
- Airway Clearance (CPT)
- TOBI
- Inhaled Steroid

PULMONARY FUNCTION TESTING

When your child is six or older, the CF team will start to perform pulmonary function tests (**"PFT's"**). This is a highly-specialized test that measures the airflow in and out of the lungs and is a good indicator of lung damage over time. Children must be able to follow careful instructions and blow out a consistent exhaled breath over a set time.

PFTs measure the "Forced Vital Capacity" (**FVC**) of the lungs—how much air you can forcefully blow out of your lungs—exhaling as hard, as fast, and as long as you can. A computer measures the first second, called the **FEV_1**, and gives back a number. We compare your child's FEV_1 number with standardized tables of what constitutes a "normal" FEV_1—based on similar traits of other children.



6 Nutrition

In this chapter:

- About pancreatic enzymes
- Why diet is important
- Roadblocks to look out for
- Handouts with more nutrition information

"I recall a day when my eight-year-old daughter, Addie, and I were traveling on the road to Seattle to see her grandparents. We were discussing the many different challenges people with different illnesses face. She mentioned that she has a friend who has diabetes and that it must be hard for her to prick herself every day. She told me about how another friend had to go home during a sleepover because her allergies caused her eyes to swell shut. Addie wished out loud that some day they would find a cure for all the diseases in the world. Just as I thought our conversation was ending, she added, 'You know what else Mommy?' 'What's that?' I asked. In a cheerful voice, Addie piped up, 'I hope when they find a cure for CF, it will be a big giant Sweet Tart!' and with a chuckle, I replied, 'I hope so too!'"

Chapter 6
Nutrition

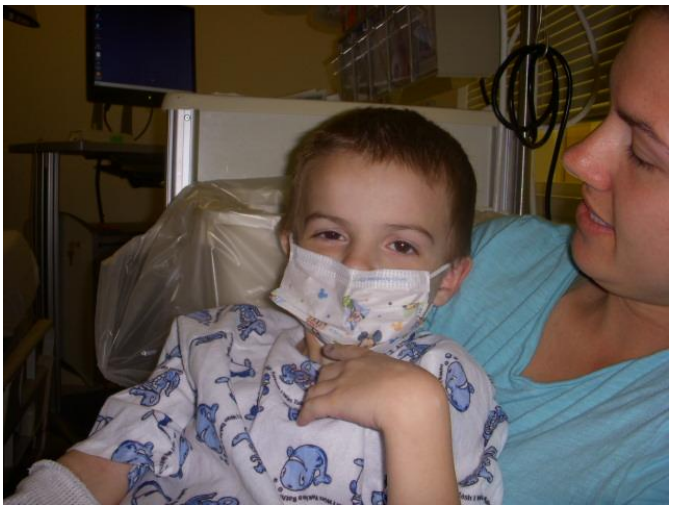
This chapter covers nutrition and the role it plays in your child's health. It seems obvious that nutrition should be prominent in a child's growth and development. However, when a child has CF, it is a bit more complicated than considering the standard Food Pyramid and three meals a day.

Pancreatic Enzymes
Your child has a disease that prevents him or her from metabolizing fat and protein in an effective way. The pancreas is often blocked with a mucus plug, in a similar way that the lungs are plugged with mucus. The function of the pancreas is to produce and distribute enzymes that help metabolize the fats and proteins that young bodies need to grow and develop. If those enzymes are not being put into the digestive system your child becomes malnourished.

One of the medications that your child's doctor will prescribe will be pancreatic enzymes. These are taken before eating and several capsules are taken at a time. Your CF team and dietician will work with you and your child to figure out the best delivery system for these enzymes. Typically, enzymes are in capsules that have little "grains" of pancreatic enzymes inside. There are many strategies for administering these "grains". You will need to know how help your child swallow the grains and eventually how to swallow pills.

Diet
Earlier we mentioned the Food Pyramid. With CF kids, most of what you know about "good" nutrition is out the window. These rules do not apply to your child. Your child has an inverted food pyramid. He or she needs a lot of fat and protein in her diet, and not so many fruits, veggies and grains. If your child is older and more verbal about their food preferences, they may prefer these other foods over the fats and proteins, simply because it is probably easier to digest. It is, however, not very effective in metabolizing and helping your child to "go and grow." Encourage the "go and grow" foods first, and then offer the strawberries and salad.

The caloric needs of your child will be great. Each age and stage your child goes through will require ever increasing calories. You now have to start thinking how to pack in the calories with extra fats and oils, and choosing foods that are rich in protein. Snacks should be intentionally high in calories. Skip the apple slices unless you intend to serve them with cheddar cheese or peanut butter.



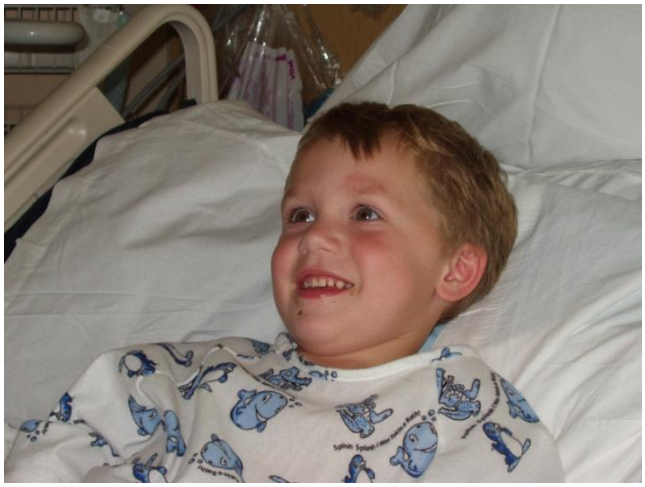
Vitamins
Your child's Pulmonologist may also prescribe vitamins for your child along with the other medications to help ensure their child's airways are clean and clear, and that pancreatic supplements are taken for digestion. The vitamins that are generally prescribed are fat-soluble; vitamins A, D, E and K. These vitamins assist in the healing and regeneration of cells. It is also a good idea for your child to take Omega 3 oils, as are found in fish oil, flax seed oil, olive oil, nuts and seeds. The Omega 3 oils help diminish the inflammatory process and help your child's cell regenerate and heal as well.

Why the fuss?
Children with CF that do not have their dietary needs met will become malnourished. Diet is huge. Your child's nutrition and their ability to gain weight and height in comparison to their peers, has a direct correlation to their lung growth and health. Healthy placement on the pediatric growth chart—for height and weight—has a direct relationship to how well your child's lungs are functioning. Therapeutic nutrition is just as important as any lung therapy your child's Pulmonologist may prescribe.

Are there any roadblocks?
You bet. Your child is going to have a lot of demands. Their regimen of pills, therapies, and trying to stay well will be compounded by the demands of making special choices in eating. Your child is going to want to express some control over all the therapies and routines that are now part of her day. There will be struggles with taking pills, enzymes and with choosing foods and eating enough. Be sure to speak with your CF Dietitian and with the others on the CF team if struggles and issues arise. They help you overcome the battles over food choices and taking all those pills.

Learning how to cook and prepare foods that will be healthy for your child will be added to your list as a parent. Enlist the help of your child's dietitian for ideas on how to cook, what to cook and what should be in your pantry. There are a lot of cooking resources for patients with CF. As your child grows older, engage them in the cooking process and get them invested in the preparation of their food. It's a great way for your child to learn about the importance of their own nutrition and may make them more willing to be a hearty eater.

You must be creative in your approach to food and eating. Many parents have battles with their children about food and eating, even when their child doesn't have CF. Because nutrition and metabolizing food is so critical to the health of your child, it will be very easy for you as a parent to fall into the trap of making meal time a battle. We often hear, "choose your battles wisely." This is especially true for you and your child now. Think carefully about how you present eating and how you model behaviors for your child surrounding the sharing of food and meals.



If your child is older and closer to the teen years it is also important to consider the power of peers and body image. The media promotes image of very thin as very beautiful. For all teenage girls this is dangerous. For teenage girls with CF, this can be especially devastating and difficult to recover from. Be aware of the signs of anorexia and talk openly with your teen about the dangers. For boys, body awareness and image may take a different tack. Boys want to

be well "toned" and "buff." Teenage boys grow fast and eat a lot. Your son with CF may be shorter, skinnier than and not as hungry as many of his peers. He may be disappointed or embarrassed about how he looks compared to his peers.

Work with your teen's friends, the CF team, school and the community to get your child active and physical. Physical activity will help both girls and boys formulate better body images and attitudes about eating and staying healthy.

7 Hospitalization

In this chapter:

- What to expect
- How to prepare
- What to bring

"My five-year-old son Conner's positive attitude never ceases to amaze me. Not only does he have CF, but he is also diagnosed with a rate disease that makes his ability to cough very weak. He spends a good part of every year either in the hospital or doing home IV's through his PICC line or taking oral antibiotics to fight his frequent lung infections.

Despite it all, he always has a smile on his face, and loves to show everyone his PICC line and G-tube to educate friends and family about his condition. He has all of his nurses wrapped around his finger and loves to meet new volunteer "friends" while in the hospital. He runs and plays with his peers and doesn't get discouraged when he can't keep up with them. Instead he just finds a different way to play that accommodates his shortfalls. I believe having these diseases makes him more compassionate towards others with illnesses; he never has complained about his diseases yet always feels for others who are sick.

No one wants his or her child to have CF, but after living it over the last five years, I wouldn't have him any other way. Conner is mature beyond his years and is so sweet. Cystic Fibrosis doesn't have him; he has CF and it's only part of who he is. In my life, it's a great honor and blessing to be his mother. He truly has taught me a lot about love, compassion, and living each day to its fullest.

Chapter 7
Hospitalization

Children with CF may be hospitalized for a variety of reasons. If hospitalization becomes necessary, good planning can alleviate some anxiety associated with the experience. Being prepared and well informed about what to expect will help you and your child. The first experience is always the hardest, and certainly not something a family looks forward to. Hospitalization can and should be viewed as a necessary part of your child's CF treatment in the overall goal of health and well being.

Preparation can make the trip easier for you and your child:

- Explain to your child that there may be a time when they will need to be hospitalized.
- Be honest with yourself, and your child, that the day will come when their health requires advanced care in a hospital setting.
- Focus on the positive outcomes that inpatient treatment can provide.
- Read age appropriate books to your child about staying at the hospital.
- Educate yourself about what procedures may be required while your child is in the hospital.
- Ask your doctor questions!

How do I check in and what will be done for my child?

- During clinic, or sometimes before if your child has been very sick, your doctor may suggest that a hospital stay is needed.
- The clinic will make the necessary arrangements to reserve a room for your child, and treatment orders should be sent to the hospital staff.
- Once you arrive, you will need to check in at the admitting counter.
- Your child will receive an ID wrist band, and after signing some paperwork, you will proceed the 9th floor.

- Check in at the nursing station, and proceed to your room.

What happens after you arrive in your room?

- A nurses assistant will take your child's vitals (temperature, blood pressure, weight)
- Some more forms will need to be signed
- Usually within the hour, an x-ray technician will show up to take an x-ray
- If the stay is scheduled for several days, a PICC line will be inserted.
- The PICC line is usually done in the child's room, or in a separate procedure room.
- A doctor will stop by to make sure everything is going well, and answer any questions.
- A menu for the day's meals should be filled out by your child.
- A hospital Chaplin will come by and explain spiritual options available.
- The CF team (nurse, doctor, dietician, social worker) will come by your room when they make their "rounds" either that afternoon, or the next morning.
- Vitals will be taken on a regular schedule, day and night!

Comforts from home to bring:
You will stay for awhile with your child. Bring some of the things from home that will be comforting for your child.

- Blanket for the bed so it looks nicer and cozier
- Special stuffed animal/toy/blanket to sleep with
- Small posters for the wall
- A few family pictures
- Favorite snacks - there is a refrigerator and microwave for families to use.
- Favorite DVD's, CD's, iPod, books, etc.
- Homework!
- PJ's and slippers

- Comfortable clothes, socks, etc.
- Toiletries, air freshener
- Anything that will make your child more comfortable and at ease

Items for Parent/Guardian:
You are here for the long haul; bring things for your entertainment as well.

- Cell phone
- A diary or log to keep track of caregivers, procedures and progress (more about why this log is important later)
- Book
- Crossword puzzles
- Sewing/knitting
- Blanket for the "guest" bed
- Snacks
- Laptop computer

How does the routine at the hospital look different from the routine at home?

- Medication will be administered via IV on a regular schedule.
- Regular treatments/other medications will be administered.
- Your child will not be stuck to the bed! Health permitting, when not receiving a dose of IV meds, your child can get up, take a walk in the halls, go to the outside courtyard, or the playroom if your doctor approves.
- Child Life staff, a great group of volunteers, will have various entertainment options and activities.
- Friends and family may visit
- A parent or guardian may stay the night in the room.

HOW TO BE A GOOD "*HOSPITAL NEIGHBOR:*"
There are lots of other kids and families in the hospital. Be mindful that everyone is trying to get well.

- Keep the noise down.
- Adhere to visiting hour guidelines.
- Talk to your nurse if you are having issues about privacy or quiet hours.
- Play rooms, family kitchens, and day rooms are shared areas, use them well but leave them tidy.
- Be aware of patient privacy and HIPAA rules. You will be notified of these rules when your child is admitted.
- If your child is in isolation, help your child and visitors adhere to these important guidelines. Hospital staff will help you understand these guidelines. (more on isolation procedures to follow).

HOW TO KEEP TRACK?

There are a lot of caregivers walking in and out of your child's room during hospitalization. Decisions about therapies and procedures may change from day to day. It can be confusing. It is very conceivable that you will be alternating nights with your child's other parent or your partner. It is important to keep track of the following bits of information so your child's care is consistent. If you can, an hourly update is a good idea to log.

- The name of your **child's nurse and CNA**
- The **doctors, residents and interns** involved in the care of your child
- Your child's **respiratory therapist**
- **Changes** in medications, therapies and dietary needs
- **Changes** in your child's symptoms
- **Keep a list of questions** to ask the nurse and the attending physicians when they come for "rounds."
- When you hand off the log to your partner, go over your notes with them. Include your child's input. This will help your child feel as though they are an important member of their care team.

Being admitted to the hospital can be a scary thought, but with a positive attitude, it can be a tolerable situation. It is extremely important to your child that you are present during their hospitalization. Your child will feel more secure and comforted. Treatments will be easier to administer if you are there to

comfort and advocate for your child. If this is a hardship on your family, please let the staff at the CF clinic know and some arrangements may possibly be made.

Please remember to **speak up** and ask questions, or ask for help dealing with issues. The CF clinic team of doctors, nurses, dietitians, and social workers are working to help your child reach their full health potential and live the best quality of life possible.

WHAT ELSE SHOULD I KNOW?

Different children with Cystic Fibrosis grow different bacteria in their lungs. Sometimes bacteria can be resistant to certain antibiotics or may present problems for other children who are also hospitalized at the same time. For this reason, some children with CF have special precautions when they are hospitalized.

The most common is "contact precautions" to prevent the spread of bacteria from one child to another. The doctors, nurses, and other staff who visit your room will wear protective gowns and gloves, and you will have some restrictions on different activities. There are policies in effect, which outline the specific restrictions; your CF care team will give you instructions when you are admitted.

Contact precautions apply to the child being admitted, as well as family members, siblings, and visitors who come and go from the room. If you are under contact precautions, ask your CF team to explain the restrictions and educate your family and visitors.

8 CF and School

In this chapter:

- Special education needs
- 504 plan information
- Laws affecting education

Chapter 8
CF and School

When your child with CF enters school, there are many factors that need to be considered. Due to increased exposure to germs, they may become ill more often and require more hospitalizations. Since there is not a lot of awareness of this disease, school staff and students need to be educated about the implications of this disease and how it may impact your child's education. Probably one of the best things that can happen is for your child to become a self-advocate. There will be questions about their size, their coughing, frequent absences, and diet. Instead of being put off by these questions, be prepared, and prepare your child with a script or a handful of resources to answer these questions. There are several resources that may help you address these concerns as they arise. Please refer to the "Resources" section to find information relevant to the schools.

Special Educational Needs

Due to their illness, your child will be eligible for either a 504 plan or an IEP (Individualized Education Plan) up to age21. These plans are designed to provide modifications/accommodations and services to your child that allow them to access the educational environment more successfully.

DIFFERENCES:

504 Plan

Definition

- Regulated under the Rehabilitation Act
- Prohibits discrimination against individuals with disabilities in programs or activities that receive federal financial assistance.
- Does not require that a disability impacts the student's ability to learn to qualify for protection under the law.

Qualifying for a 504 Plan

- Individuals with a physical or mental impairment that substantially limits one or more major life activities (attendance, extracurricular activities, field trips, homework, school camps, physical education)
- A person with a record of an impairment (documentation is required from your physician)
- A person who is regarded as having such an impairment (chronic cough, G-tube, frequent absences, dietary needs)

IEP/IFSP (Individualized Education Plan/Individual and Family Service Plan)

Definition:

- Mandates that all eligible children are entitled to a **FREE AND APPROPRIATE** public education.
- Related services may be provided for the child to benefit from their educational experience. These can include: transportation, health services, administration of medications, modifications/accommodations in education.

- An IFSP serves children ages 0-5.
- An IEP serves students ages 5-21.

Qualifying for an IFSP/IEP:

- A student may be considered "other health impaired" and, therefore, eligible for services under the IDEA (Individuals with Disabilities Education Act).
- Cystic Fibrosis impacts the ability for the child to learn due to absences or requiring a school to adhere to medical and therapeutic treatment during the school day.

(SEE TABLES BELOW FOR MORE DETAIL/CLARIFICATION ON LAWS AND ELIGIBILITY)

Both IFSP/IEP and 504 Plans consist of modifications and/or accommodations to education. If any of these apply to your child, they will qualify for services under one of these plans. Examples of modifications/accommodations for children with CF may include the following:

- Allot time during the school day for the student to receive medical treatments, physical therapy or to rest.
- Modify school rules for student self-administration of medication such as pancreatic enzymes.
- Allow students to eat snacks/have access to water more frequently during the school day.
- Modify physical education requirements.
- Establish a plan for homework when student is absent, including extended time for tests, assignments and grading.
- Modification of workload, homework, and tests to accommodate heath status, fatigue and absences.
- Homework sent home after one absence.
- Preferential seating/classroom assignment (away from ill students or another student with CF).
- Exemption from the attendance/tardiness policy.
- Tube feedings in school or feeding therapy.
- Full-time nurse in building at all times or a person trained in medical protocol (feeding, IV therapy, chest therapy, changing G-tubes, administering medications).
- Air-conditioned classrooms
- Handicapped parking
- Transportation to and from school

- Full participation in extracurricular activities and field trips.
- Home or hospital instruction after three absences.
- Provide audio or video recordings of classes missed.
- Provide teacher assistance for full academic load (tutoring).
- Second set of textbooks at home and/or separate set of supplies at school to be used solely for child at school (for infection control).
- Established plan to manage medical emergencies at school.
- Unlimited access to private bathroom at school (such as a nurse's office).
- Unlimited access to hand washing and/or sanitizer.

SEE ATTACHED EXAMPLE OF WORKSHEET FOR DEVELOPING A 504 PLAN OR GENERATING MODIFICATIONS/ACCOMMODATIONS TO THE IFSP/IEP.

LAWS AFFECTING EDUCATION

Individuals With Disabilities Education Act (IDEA)	Federal law that requires public schools to accommodate children with special health or educational needs. **This is for an IFSP/IEP.**
Rehabilitation Act of 1973, Section 504	Federal law that prohibits discrimination against an individual solely because of a disability. This law applies to public schools and private schools receiving federal funds. **This is for a 504 Plan.**
Americans With Disabilities Act (ADA)	Federal law that prohibits discrimination against an individual because of a disability in a variety of settings, including schools.
State Education Laws	Some states have laws that protect students regarding the self-administration of medications, such as digestive enzymes. Some states have laws that restrict who in the school can administer medication to a student.

TIMELINE FOR THE IEP/IFSP PROCESS

Identification	Inform the school of need for evaluation for IDEA services to a child's teacher, principal, nurse, psychologist, or learning specialist.

Evaluation	An evaluation is done by the school and must be completed within **60 calendar days** of the request.. The school will need to obtain a physician's statement from your child's doctor.
Team Meeting	The school must give parents at least five days notice of a scheduled meeting. The initial meeting must be held within **30 days** of the completion of the initial evaluation. Most schools hold team meetings at the beginning of the school year.
IFSP/IEP	The school must send parents a copy of the drafted IFSP/IEP within **30 days** of the initial team meeting. Parents also must receive a copy of their parental rights and all IFSP/IEP documentation for their personal records. This includes evaluation reports, physician statements, eligibility form, IFSP/IEP document, permission to evaluate, team meeting notice.
Approval and Acceptance of IFSP/IEP	Parents have **30 days** from receipt of the IFSP/IEP to accept, partially accept, or deny the plan. An accepted plan (or accepted portions of a plan) should be implemented immediately after parents sign and return the IFSP/IEP.
Ongoing Review	The team should meet **AT LEAST ONCE A YEAR** to review the IEP and **EVERY SIX MONTHS** to review the IFSP.

For more information regarding specific laws that affect you in your state, please refer to the following website: http://www.actioncf.org/school.html

What to expect in this process:

- YOU need to be a strong advocate for your child.
- Ask questions.
- Make certain the procedures are being followed.
- Be certain that the school is in compliance.
- Understand the laws.
- DON'T be intimidated by the process.
- Ask all the questions you need to until you understand.

Having a child who qualifies under the "other health impaired" category is rare. Therefore, qualifications for this eligibility may be overlooked. Typically, this eligibility is used for children who qualify as having ADD/ADHD where the impact on learning is clearly identifiable. If a school does not understand why students with CF should be considered "other health impaired," explain the limitations the disease places on these students in relation to their ability to attend school regularly and their need for administering medical treatments during the school day.

If you have difficulty obtaining an evaluation from the school, contact the State Education Agency to file a complaint.

Oregon: (503) 378 - 3569

Washington: (360) 725 - 6000

You may need to request the presence of an advocate for your child and family. THIS IS YOUR RIGHT! Advocates may include: a doctor, nurse, social worker, an attorney, a representative from the Cystic Fibrosis Family Councilor you can contact a representative from a parent advocacy group.

The school district may be reluctant or "drag their feet" when you request developing a plan for your child (particularly a 504 Plan). YOU will need to be persistent in assuring that a meeting is scheduled in a timely manner. It is advisable to document **all** contact with the school. This includes phone calls, written letters, e-mail correspondence, personal conversations, etc.

Once the documents and plan are in place, be sure to check in frequently to assure that the school is compliant. This becomes particularly important in secondary school when your child has multiple teachers/classes, a higher intensity of homework, and their social life is amplified.

9 Financial Impact

In this chapter:

- Private health insurance
- State health insurance
- Assistance programs

Chapter 9
Financial Impact

Cystic Fibrosis is a condition that impacts all aspects of your finances. Because medical care and medications for children with Cystic Fibrosis are very expensive, you must have medical insurance coverage. Having good health insurance for your child is extremely important. It is also important that you become knowledgeable about resources and where you can get assistance.

Health Insurance
Choosing a policy or understanding your policy can be difficult and it is easy to get overwhelmed.

It is very important to:

- Obtain a copy of your policy from your employee benefits or call your insurance company.
- Make sure you read and understand your insurance policy
- Carefully go over the sections important to CF care, especially prescription coverage. Even the best insurance can be costly with premiums and co-pays

The best person to help you advocate, especially if problems arise, is the case manager at the insurance company. Ask your insurance company for the case manager and once you meet them, make them your new friend.

Your insurance might not pay some medical bills at first, so always start with your case manager. If you receive a denial of coverage for a medication or treatment, you have the option to appeal.

Changes with Insurance
There are federal and state laws that affect health insurance. The 'Consolidated Omnibus Budget Reconciliation Act' (COBRA) can help you keep your group insurance if you lose your job, but you will have to pay the monthly premiums. COBRA is often used to avoid a gap in between insurances and coverage can be very expensive.

Most health insurance plans have a specific period of time where a pre-existing medical condition (CF) is not covered. It is important to know about called Creditable Coverage, which reduces the amount of time your new insurance can limit coverage based on a pre-existing condition, meaning that you get credit for each month you had coverage under your previous plan. This applies if you have not had more than **63 days** without insurance. You can get a statement of Creditable Coverage from the insurance company that is ending and give it to the new one.

State Health Insurance
If you know that your child will be without any insurance, it is worth the time and effort to be informed about the State offerings. There might be a program your child qualifies for. The following is a brief description and overview of the State Assistance Programs.

There are high-risk health insurance pools offered in Oregon and Washington. These programs are designed to cover children that are denied coverage because of pre-existing medical conditions, such as CF especially when COBRA benefits have been exhausted and there are no other insurance options.

These programs have plans you may choose from and may require a paid premium, deductibles and co-pays.

Oregon Medical Insurance Pool (OMIP)
1-800-848-7280 www.omip.state.or.us

Washington State Health Insurance Program (WSHIP)
1-800-877-5187 www.wship.org

Both States offer family health insurance assistance programs. The programs assist in providing insurance premium subsidies for low-to-moderate income families who are otherwise uninsured and meet other eligibility requirements. The amount paid depends on the family income and number of family members.

Oregon: Family Health Insurance Assistance Program (FHIAP)
1-888-564-9669 www.fhiap.oregon.gov

Washington: State Children's Health Insurance Program (SCHIP)
1-877-543-7669 www.dshs.wa.gov

If you qualify as having low income, both States have Medicaid Programs:
The Oregon Health Plan has several different plans assigned by the county you live in. Some of the local plans require a referral or authorization for CF clinic visits and medications.

Oregon Health Plan (OHP) 1-800-359-9517

Washington Medicaid is assigned per plan; some are more restrictive about access to the CF clinic.

Washington Medicaid 1-800-204-6429

Social Security Benefits
Social Security also known as Supplemental Security Benefits or SSI for children has strict medical and non-medical requirements. This means that even though your child has been diagnosed with CF, your child does not automatically qualify for social security. If your child meets the medical requirements, your family income has to meet the low income criteria.
If you decide to apply, the CF Clinic has an Advocacy Manual with information on how the SSI application process works.

Social Security 1-800-772-1213 www.ssa.gov

Medication Assistance Programs
Medication assistance programs vary based on the type of drug and manufacturer. Some help with high co-pays; others will only help if you have no prescription coverage. The CF Clinic or your pharmacy should be able to inform you of what programs are available for what medications. There is usually an application process for each prescription. Some programs are income based and some are not.

OHSU Financial Assistance
Request for financial assistance may be made at any point before, during, or after the receiving care. OHSU offers an application process for determining eligibility for financial assistance. You just need to ask.

Financial assistance is specific to each patient admission and is granted for medical procedures. Financial assistance related to outpatient services will require periodic screening for changes in eligibility.

If paying your medical bills becomes a financial burden, you should apply for the Statement of Financial Resources (SFR). It is based on income with discounts adjusted according to income and household size. The SFR is assessed yearly.
You can request an SFR form from Registration or Patient Billing.

Registration 503-494-8505
Patient Billing 503-494-8760 and 503-494-8417

The Financial & Medicaid Services at OHSU can assist you in determining if your child qualifies for the Oregon Health Plan (OHP). They can help with the application process. They also have information on Washington Medicaid programs.

OHSU Financial & Medicaid Services 503-494-1671

The CF Clinic - Social Work
Please contact the CF Clinic when you have problematic financial concerns. Other funds and grants for financial assistance may be available depending upon your situation and the type of assistance needed. Your Social Worker can help you. Let them know as soon as possible when you know that there might be changes in your financial situation.

Oregon Center for Children with Special Health Needs (OSCSHN)
This is a state program that will cover medical assistance based on 'growth and development.' Generally, assistance is limited to nutritional supplements and some medical equipment.

Tax Implications
There are many parts of CF care that may be deductible or improve your tax burden. You are encouraged to consult with a licensed tax preparer to check for a tax exemption credits on State and/or Federal forms for medical expenditures.

It is worth reviewing eligibility for laws that guide possible tax deductions. An Internal Revenue Service representative at the local branch office should be able to assist you in understanding them. For example, you receive an additional exemption if your child with CF has an Individualized Educational Plan (IEP) in place.

Additional Information
Another resource that can be helpful to answer your financial questions is:

CF Legal Information Hotline 1-800-622-0385

10 Research and Drug Trials

In this chapter:

- OHSU's participation
- Drug trial stages defined by phase
- Other research

If I Had 100
by Robert, Age 8

"If I had 100 pencils I would open a school supply store.

If I had 100 mosquito bites I would be very itchy.

If I had 100 friends I would play all day.

If I had 100 legs I would scare people.

If I had 100 eyes I would have a really big head.

If I had 100 geckos I would need a lot of crickets.

If I had 100 dollars I would donate it to the CF Foundation.

If I had 100 pieces of cheese I would need a lot of crackers.

If I had 100 cures for CF, I would cure me and 99 other people."

Chapter 10
Cystic Fibrosis Research and Drug Trials

When he Cystic Fibrosis Foundation (CFF) was founded in 1955, it set high goals to advance understanding of CF, to develop new treatments, and to find a cure. As a result, many new treatments have been developed, and many more are in the works, forming a "pipeline" of potential new CF therapies. There are currently dozens of therapies in development—more than at any time in the CFF's history.

To maintain this pipeline with promising treatments, many CF researchers continue to translate new knowledge from basic science laboratories into potential therapies.

The Oregon Health and Science University CF Clinic is accredited by the Cystic Fibrosis Foundation and has participated—and continues to participate—in many trials for new CF treatments.

DRUG TRIALS

One of the most important areas of research is looking into new drugs to treat CF. The process starts with an idea: a compound developed in a laboratory may look promising, perhaps an antibiotic that fights lung bacteria, or a chemical that blocks inflammation. But getting from the idea stage to actually having a marketable drug that your doctor can prescribe is a difficult, lengthy, and expensive process.

Because CF is a "cascade" of events within the body, drugs are developed to attack the disease at different points along the chain. An example is illustrated below:

Cascade of Events	***Treatments***
Defective Gene	*Gene therapy (still in early stages of development)*
V	
Defective CFTR	
Abnormal Ion Transport	*Aactivating alternate chloride channels (i.e. Denufosol)*
V	
Bronchial obstruction	
Infection	*Inhaled antibiotics (i.e. TOBI, Aztreonam)*
V	
Inflammation	
Tissue destruction	*Anti-inflammatories (i.e. azithromycin, ibuprofen)*

Drug trials go through phases (summarized below). Lots of testing and documentation occurs even before drugs get to the first stage; it usually takes several years to get FDA approval for a new drug once it is developed.

The OHSU CF Clinic may have different study drugs in different phases going at any particular time, though we usually participate in Phase II and Phase III studies.

Clinical Trial Stages

Phase I
- Lasts several months
- Not looking at efficacy, just if drug is safe or not
- Analyzing metabolism and side effects
- Small number of participants

Phase II
- Looking at efficacy: How well drug works in humans
- Usually study vs. control groups—moderate number of participants

Phase III
- Concentrates on drugs with favorable earlier trials
- Large number of subjects
- Determine appropriate dosage
- Safety and effectiveness
- FDA approval for therapy

Phase IV
- May be required
- Long-term safety and effectiveness

Participation in drug trials is an extremely important. Without human testing, drugs to benefit CF patients would not make it to the market and key therapies that benefit CF patients would not be available. All research is carefully monitored by both the FDA and an Institutional Review Board. Each trial is meticulously designed to be as safe as possible for the participants.

For each trial, the CF Clinic contacts potentially eligible patients by telephone or mail, in order to recruit participants that will give us good information about the drug. All participation is completely optional and your child would never be enrolled in a trial without your permission. You may choose not to participate in drug trials, or to drop out of one at any time, and doing so would never impact the quality of care you receive from the CF Clinic staff.

OTHER RESEARCH

There are other areas of research that the OHSU CF Clinic participates in. All patients are asked if they would be willing to participate in the CF Foundation Patient Registry, which collects epidemiological data on all of the patients we see—things like lung function tests, height/weight, and medications. The registry collects this information in a huge national database. It's the oldest and largest database of its kind in the country and has proven invaluable for developing new CF therapies. The information is accessed by researchers, who can perform complex statistical analyses—for example how well medications might be working, or how to better improve dietary regimens. The best part is, the registry only requires your signature for permission; we do all of the data collection "behind the scenes" and of course, all information is confidential and anonymous. No one will ever use the database to sell you products or try and get your personal information.

From time to time, OHSU participates in Quality Improvement initiatives, designed to improve our flow, delivery of care, and how well patients are treated. You may be asked to participate in this kind of research, which often includes questionnaires and interviews. We use this information to improve our quality of care.

Research is a key component to fighting Cystic Fibrosis. Working together, we can come up with drugs, treatments, and plans to defeat CF.

If you hear of a trial or research protocol you'd be interested in learning more about, or even participating in, please contact the center nurse. Chances are we've heard of the research, or may be looking for patients to enroll in the trial.

For more information, please visit http://www.cff.org and click on the "research" menu item.

11 The CF Foundation

In this chapter:

- CF foundation facts
- Special programs
- Contact information

"When our daughter was first diagnosed with Cystic Fibrosis I remember being overwhelmed by the negative picture from all the information we initially found. CF long-term prognosis was not good and the shortened life expectancy was heartbreaking. But as we dug deeper and investigated more we began to see hope and promise of a better future than even ten years ago. We began to see that through the Cystic Fibrosis Foundation amazing research advancements were being made every day. The future was looking better and better. Obviously we wish CF was not a part or our daughter's life, but at least she is living with it now—a time when there is not only hope but promise and commitment grounded in cutting-edge research. There are new antibiotics, new nebulized medicines, and new vitamins developed just in the four years since our daughter was diagnosed. We have found a fantastic partner in the CF Foundation and it has become a source of comfort knowing we have then fighting alongside us every step of the way."

Chapter 11
Cystic Fibrosis Foundation

The mission of the Cystic Fibrosis Foundation, a **nonprofit** donor-supported organization, is to assure the development of the means to cure and control Cystic Fibrosis and to improve the quality of life for those with the disease.

Foundation Facts

- Founded in 1955 by parents of CF patients who wanted an avenue for fundraising and research.
- The Foundation is the leading organization in the United States devoted to Cystic Fibrosis.
- In 1989, CF Foundation-supported scientists discovered the defective gene that causes Cystic Fibrosis—a monumental breakthrough on the road to a cure.
- Funds and accredits more than 115 **CF care centers,** 95 adult care programs and 50 affiliate programs
- Has 80 **chapters and branch offices** nationwide.

The Foundation is Involved

- Investors funding drug development and discovery
- Scientists researching new avenues of treatment and care
- Advocates keeping CF a top priority in government, industry and research
- **Fund-raisers** securing the money needed to support their efforts
- Resources for support, information and links to specialized CF care

Volunteer Organization

- The Foundation receives no federal funding, so depends on the generosity of individual donors, corporations and foundations.
- More than 250,000 dedicated volunteers devote their time and talents to help raise funds for research and medical programs. They are the engine of the Foundation and make progress possible.
- In 1980: raised $40 million
- In 2008:projected to raise $100.6 million
- By 2010:needs $121.4 million to continue funding programs and research
- The Foundation's 80 chapters and branch offices host thousands of special fund-raising events year-round, such as golf tournaments, celebrity concerts, and dinner dances.

Local events - Contact the CF Foundation for event dates

- Chef's Dinner & Wine Extraordinaire
- Annual Great Strides Walk
- Wine Opener
- Bob Gilder's Platt Golf Challenge
- Wachovia Annual Golf Tournament
- Unmask the Cure Gala Event
- Texas Hold 'Em Poker Tournament
- CureFinders

SPECIAL PROGRAMS

Cystic Fibrosis Services Pharmacy

- Established in 1988 as a wholly owned subsidiary of the Cystic Fibrosis Foundation.
- A specialty pharmacy to provide availability and access to CF medications, as well as assistance with insurance issues faced in obtaining CF medications.
- Provides personalized service, patient advocacy, patient education and reimbursement support to the CF community.
- CF Services is a full-service pharmacy and carries an extensive line of products, including but not limited to:
    - pancreatic enzymes
    - nebulized medications and supplies
    - inhalers
    - antibiotic medications
    - transplant medications
    - diabetes medications and supplies
    - nutritional items
    - over-the-counter items
    - and a variety of other prescription medications

CF Legal Information Hotline

Provides free information about the laws that protect the rights of individuals with Cystic Fibrosis. It serves as a resource for CF Care Centers, individuals with CF and their families. Information available:

- Obtaining health insurance
- Access to health insurance under the Health Insurance Portability and Accountability Act (HIPAA)
- Extending health insurance coverage under COBRA
- Insurance coverage issues
- Social Security Disability Insurance benefits
- Supplemental Security Income (SSI) benefits
- Medicare and Medicaid coverage
- Employment issues for people with CF, including protections under the Americans with Disabilities Act (ADA)
- School laws to assist children with CF in public elementary and secondary schools through the Individuals with Disabilities Education Act (IDEA)
- Protections for students in elementary school through college and beyond under the Rehabilitation Act of 1973, Section 504
- Unpaid time off from work under the Family Medical Leave Act (FMLA)

The CF Legal Information Hotline is an information resource only. The Hotline does not provide legal representation for callers. All calls are confidential.

CYSTIC FIBROSIS FOUNDATION CONTACTS

Oregon Chapter
9320 SW Barbur Blvd. Ste. 210
Portland, OR 97219
(503) 226-3435 / (800) 448-8404
email: oregon@cff.org
www.cff.org

Cystic Fibrosis Services Pharmacy Hotline
(800) 541-4959
www.CFServicesPharmacy.com

Cystic Fibrosis Legal Information

(800) 541-4959
email: CFLegal@cff.org

12 CF Support and Information Resources

In this chapter:

- Websites
- Books and recommended reading
- Checklists
- Example help sheets

Chapter 12
CF Support and Information Resources

We are happy to provide this handbook for you and this chapter is a way to supplement and extend the information found in other chapters. Please explore the websites and read the books, but remember they won't all be perfect for you. Many of the websites here (and others you will find) will have chat rooms and forums for community discussions, and as with all other similar forums there are people out there trying to help and support the others reading and participating, but at the same time there are people out there with very negative perspectives. Just be aware that not everyone posting is really trying to be helpful to you, but many people just want to share their experiences with others and hope that they can help and be helped by these community forums.

We have arranged this list of resources by subject, then by media and target audience. Some websites may be listed in multiple locations. Explore, learn and ask questions; we are at a time when the more you know, the more you will understand how much hope there is for children and adults with Cystic Fibrosis.

Information about Cystic Fibrosis

Website Name	**Web Address**
Cystic Fibrosis Foundation	www.cff.org
Cystic Fibrosis Research, Inc	www.cfri.org
Cystic Fibrosis Center at Stanford	cfcenter.stanford.edu
National Library of Medicine and National Institute of Health	www.nlm.nih.gov/medlineplus/cysticfibrosis.html
KidsHealth for Parents	www.kidshealth.org/parent/medical/lungs/cf.html
CF Voice	www.cfvoice.com

Book Title	**Author**	**Target Age**
Mallory's 65 Roses	Diane Shader Smith	Children 4-10 yrs.
CF and Me	Susan Tumiel Smith, M.Ed.	Children 3-6 yrs.
Big Pats or Little Pats?	Susan Tumiel Smith, M.Ed.	Children 1-3 yrs.
Cystic Fibrosis: The Ultimate Teen Guide	Melanie Ann Apel	Teens and Parents

Living With CF

Website Name	Web Address	Target Audience
Cystic Fibrosis Foundation	www.cff.org	All
CF Voice	www.cfvoice.com	All
Cystic Fibrosis.com – Online support	www.cysticfibrosis.com	All

Book Title	Author	Target
Parenting Children with Health Issues	Foster W. Cline MD and Lisa C Greene	Parents and Caregivers
Not to Worry, Mom, I'm Okay: Lessons in Living from a Beloved Son	Karma Smith Belnap	Parents and Family
Cystic Fibrosis: A Guide for Patient and Family	David M Orenstein	All
Bittersweet Chances: A Personal Journey of Living and Learning in the Face of Illness	Dana Selenke Broehl	Parents, Friends and Family

Nutrition and CF

Website Name	Web Address
Kids Health	http://kidshealth.org/parent/nutrition_fit/nutrition/cf_nutrition.html
Nutrition and CF	http://cf.conncoll.edu/nutrition.html

Book Title	Author
Fat and Loving It	Gail Farmer
A Way of Life: Cystic Fibrosis Nutrition Handbook and Cookbook	Mary Marcus, R.D., et al. UW (Wisconsin) Hospital and Clinics Order at: http://www.lulu.com/content/2482231
Pass the Calories, Please!	Gail Farmer
Cruising on Next Stop…Adulthood	Digestive Care, Inc., CF Foundation Education

CF Stories

True and fictionalized stories of life experiences with CF

Title	Author
The Spirit of Lo (see www.spiritoflo.com)	Terry and Don Detrich
Alex: The Life of a Child	Frank Deford
A Little Love Story	Roland Merullo
The Power of Two: A Twin Triumph over CF	Isabel Stenzel Byrnes; Anabel Stenzel
Robyn's Book	Robyn Miller

GLOSSARY

Glossary

A

ABSORB/ABSORPTION
The passing of *nutrients* into the blood stream. This occurs mainly in the small intestine after food is *digested*. Nutrients that are absorbed include proteins, fats, and carbohydrates. Vitamins and minerals are also absorbed.

ACBT (Active Cycle of Breathing Technique)
A technique to help clear secretions from the chest and lungs using different types of breaths. See *huff coughing.*

ACUTE
A sudden episode of disease or illness that only lasts a short time. In CF, a chronic disease, acute *infections* are common.

AEROSOL
Particles of a solid liquid forming a mist in the air. In breathing treatments, an air compressor forces medicines to form a mist. Then, they can be inhaled directly into the lungs.

AIRWAYS
The tubes that carry air in and out of the lungs. These tubes begin with the nose and mouth and include the trachea (windpipe), bronchi, and bronchioles.

ALLERGENS
The things to which people are allergic. Common allergens are dust, pollens, and molds.

ALLERGY
The body's overreaction to a substance, an *allergen*. This occurs when the body has become sensitive to that substance. Allergic reactions may include rash, wheezing, airway or sinus problems, or hives.

ALVEOLI
Tiny air sacs at the ends of the airways, deep in the lungs, where the body gets its fresh oxygen supply. The walls of the alveoli are covered with tiny blood vessels. As blood is pumped through these vessels, oxygen passes from the alveoli into the bloodstream. At the same time, carbon dioxide passes out of the blood into the air sacs. With each breath, we *exhale* carbon dioxide and *inhale* oxygen-rich air. A single air sac is also called an "alveolus."

ANTIBIOTICS
Medicines used to fight infections caused by *bacteria*. They kill bacteria or prevent them from growing. Antibiotics may be given by mouth, by injection, or by IV (intravenously). During breathing treatments, antibiotics may be *inhaled*.

ASTHMA
A medical condition in which the airways overreact to stimulation. Severe episodes or attacks of asthma may be triggered by *allergens*, exercise, infections, or smoke. Different people are affected by different triggers. These reactions cause the airways to become narrower, which makes breathing harder. The symptoms of asthma may include *coughing, wheezing,* and shortness of breath. Medicines may be given to prevent the overreactions of asthma and treat acute episodes.

AUTOGENIC DRAINGAGE (AD)
A type of *chest physical therapy* a person with CF can do alone (it means "self-drainage") It uses varying patterns of breathing and *huff coughing* to help get *mucus* out of the lungs. Autogenic drainage can be done in any position that is comfortable; gravity is not used to move the mucus. The CF team teaches the proper techniques. Autogenic drainage requires greater body awareness than other forms of chest physical therapy, and is usually taught to patients who are at least twelve years old.

AUTOSOMAL RECESSIVE GENE
One of the several ways that a trait, disorder or disease is passed down through families. Two copies of the gene must be present to develop the trait, disorder or disease.

B

BACTERIA
Living organisms that may cause infections. Most people with CF get bacterial lung infections, which are treated with *antibiotics*. Our bodies need some bacteria to function. These normally live and grow in the body without causing harm.

BASELINE
One's normal state of health. The baseline or usual level of functioning includes a number of considerations, such as the amount of coughing, exercise tolerance, and breathing effort. One's baseline health is often referred to for comparison. For example, after a pulmonary exacerbation, the goal of treatment is to return someone to his or her baseline state of health.

BLOCKAGE
When something blocks a passage. In the lungs, a blockage prevents air from moving freely in and out of the lungs (or part of a lung). Any solid or semisolid substance may cause a blockage. In people with CF, *mucus* may block airways. Blockage may also occur in the intestine.

BLOOD GAS
The level of oxygen and carbon dioxide (CO_2) in the bloodstream, especially in the arteries. The term "blood gas" is also used to refer to the test and the actual measurement of oxygen and carbon dioxide.

BRONCHI
The large airways in the lung. The trachea (windpipe) branches into two bronchi, which carry air into each of the two lungs. Each bronchus (singular of bronchi) then branches into many smaller *bronchioles*.

BRONCHIECTASIS
A malformation of the bronchiole tubes. In CF, this results from chronic infection and inflammation.
BRONCHIOLE

The smallest of the airways in the lungs. The trachea (windpipe) branches into large airways called bronchi. In turn, these branch out into many bronchioles. The bronchioles reach deep into lung tissue and end in *alveoli*. The airways also carry air to and from the alveoli.

BRONCHITIS
An inflammation of the lining of the *bronchi* (airways).

BRONCHODILATOR
A type of medicine that relaxes the airway muscles. This allows the airways to open wider. In CF, bronchodilators are used to help patients breathe more easily. The medicines are particularly helpful in treating *asthma*.

BRONCHOSPASM
The tightening of the muscles in the airways. This causes the airways to narrow and may make breathing harder. Bronchospasm often occurs in *asthma*. It may cause *coughing, wheezing,* and shortness of breath.

BURKHOLDERIA
A group of bacteria that can colonize the lungs of patients with CF. These bacteria are more resistant to standard therapy.

C

CALORIE
A measure of a the amount of energy the body gets from food. People with CF may need more calories than other persons. Their bodies need extra energy to fight infection and cope with lung damage. People with CF also need to eat more if they do not *absorb* all the food they eat.

CARRIER
A person who has a single *gene* for an *autosomal recessive* genetic disease, such as CF. A carrier has one gene for the disease and one normal gene. Carriers do not have the disease, but they may pass the gene onto their children.

CELLS
The basic units of all living things. The human body has many different types of cells. CF affects the cells lining the *exocrine glands, causing them to not work normally.*

CENTRAL LINE
An *IV* or catheter placed in a large chest vein. A central line may be needed when intravenous (IV) medicines or nutrition are given frequently. Some *antibiotics* or total parental nutrition (*TPN*) may be given by the central line. The catheter's opening is usually in the neck upper part of the arm. A brief procedure is needed to start a central line.

CENTRAL PORT
Refers to a device for intravenous access in patients who require frequent or continuous administration of intravenous substances.

CF
Acronym for Cystic Fibrosis.

CF PANEL
Blood work drawn in the laboratory to check for CF-related complications. Usually done annually.

CPT (CHEST PHYSIOTHERAPY)
Physical methods used to loosen and help clear *mucus* from the lungs. CPT may include *postural drainage, percussion, vibration,* and exercise. Other kinds of CPT include *autogenic drainage* and *ACBT*.

CHROMOSOMES
The parts of the *cell* that carry *genes*. Chromosomes are found in every cell of the body. Normally, a person has 23 pairs of chromosomes (a total of 46 chromosomes). When is child conceived, each parent provides half the chromosomes, one for each of the 23 pairs. Each chromosome may contain hundreds of genes. The CF gene is carried on the seventh chromosome.

CHRONIC
A condition that lasts a long time. Most chronic diseases can't be cured. Treatments are used to manage the *symptoms* and effects of the disease. *Acute* episodes or illnesses may occur as part of a chronic disease. CF is a chronic disease.

CILIA
The tiny hair-like structures lining the cells of the large airways. Cilia help move mucus and foreign matter, such as dust and *bacteria,* up and out of the lungs.

CLEANOUT
Occasionally CF patients check-in to the hospital for getting a cleanout. The purpose is to rid the lungs of excessive infections and mucus. Treatments include intravenous antibiotics, chest physical therapy, and rest.

CLUBBING
A condition in which the ends of the fingers and toes are enlarged or bulblike. The nails may become rounded. Clubbed fingers and toes sometimes appear in people who have a chronic lung disease; such as CF. The amount of clubbing isn't always related to the severity of the illness. The actual cause of clubbing is unknown.

COMPLICATION
A problem caused by an underlying disease. Common complications of CF are lung infection, DIOS), and sterility.

COMPRESSOR
A device used along with a nebulizer cup to administer lung treatment medications such as Albuterol or dNase.

COUGH
The quick and forceful exhalation of breath out of the lungs. Coughing helps to clear the airways of foreign matter and mucus.

CRACKLES
A sound indicating an airway blocked by mucus, heard by the CF doctor with a stethoscope.

CULTURE
A process of growing bacteria taken from a patient, in the lab. Culture techniques are used to determine sensitivity to antibiotics.

CYSTIC FIBROSIS (CF)
A hereditary (genetic) disease that affects the lungs, digestive system, sweat glands, and male reproductive organs.

D

DIABETES
A disease in which the body loses the ability to make insulin. Insulin is produced by the *pancreas* and controls how much sugar the body can use for energy and how much is stored. Without insulin, people cannot get the energy they need from the food they eat. Diabetes is usually treated with daily insulin shots and a carefully controlled diet.

DIAPHRAGM
The main breathing muscle, a dome shaped muscle separating the chest and abdomen. People with CF may learn to use their diaphragms to make breathing easier. To do this, they work with physical therapists or respiratory therapists.

DIGEST/DIGESTION
The process of breaking down food into *nutrients* and *absorbed* into the blood stream and used by the body. Digestion begins in the mouth and continues in the stomach and small intestine.

DIGESTIVE SYSTEM
The mouth, esophagus, stomach, small intestine, liver, *pancreas*, large intestine, and anus. These parts of the body provide food or nutrition for the whole body. They take in food and then *digest* the food and remove the wastes.

DIOS (Distal Intestinal Obstruction Syndrome)
Involves blockage of the intestines by thickened stool. Symptoms include crampy abdominal pain, vomiting, and a palpable mass in the abdomen. Surgery is often required to relieve the obstruction if enemas and laxatives don't work. Individuals prone to DIOS tend to be at risk for repeated episodes. Ongoing treatment includes pancreatic enzyme replacement and stool softeners.

DNA (Deoxyribonucleic Acid)
The basic building blocks of the body's structure and function.

DNASE (Deoxyribonuclease)
A medicine that that helps make thick mucus thinner.

DRUG LEVEL
A lab test to measure the amount of a medicine or drug in the blood. To check a drug level, a blood sample is taken and sent to the lab. A drug level shows whether or not the correct amount of medicine is in the blood. If not, the dosage of medicine is adjusted. Drug levels may be need for some medicines.

E

EDEMA
A condition in which fluid building up in tissues causes swelling or congestion. Edema may show up as swollen feet or ankles. It has several causes. When it occurs in the lungs as a result of heart failure, it is called pulmonary edema.

EMBOLIZATION
A procedure to stop bleeding in the lungs. A long thin tube is threaded to the blood vessels that lead to the *bronchi*. Little plugs of plastic-like material are then injected through the tube to block the leaky blood vessel. Bleeding from the lungs is rare and uncommon.

ENZYMES, Pancreatic
Chemicals the body uses to break down or *digest* food. Without them, food won't be broken down into *nutrients* the body can use. The *pancreas* produces enzymes that digest food. In CF, *mucus* may block the ducts or tubes that carry enzymes in the pancreas. As a result, the pancreatic enzymes can't reach the small intestine, and food will pass through the intestinewithout being digested. Most people with CF must take pancreatic enzymes to digest food.

ESOPHAGITIS
An inflammation or irritation of the esophagus, which may be caused by *gastro-esophageal reflux* (GER). Esophagitis may cause heartburn (pain in the chest).

ESOPHAGUS
The tube that connects the mouth to the stomach.

EXPIRATION
Breathing out; the flow of air out of the lungs with each breath.

EXOCRINE GLANDS
Tissues in the body that make secretions. The sweat, salivary, and tear glands are exocrine glands. In the airways and *pancreas*, exocrine glands produce *mucus*, or they may produce abnormal secretions.

F

FIBROSIS
Scarring.

G

GASTRO-ESOPHAGEAL REFLUX (GERD)
The flow of stomach contents back up into the *esophagus*.. Gastroesophageal reflux may cause vomiting, heart burn, and *esophagitis*. Because of GERD, a person may aspirate or *inhale* stomach contents into the lungs. This may cause *pneumonia* as a *complication* of GERD. Medical and surgical treatments for GERD include medications and *fundoplication.*

G-TUBE (Gastrostomy Tube)
A gastrostomy feeding tube insertion is the placement of a feeding tube through the skin and the stomach wall, directly into the stomach.

GENE
The basic unit of heredity. Genes determine each person's physical and chemical makeup. They are carried on as *chromosomes*. Each of a person's 46 chromosomes carries hundreds of genes. CF is caused by an abnormal gene located on the seventh chromosome.

GENETIC
Anything related to genes. CF is a genetic disease. Parents may carry a gene in their chromosomes that causes CF and may pass that gene on to their children.

GENETIC COUNSELING
A discussion with a medical professional trained in genetics. The counseling may help a couple determine their risk of passing on an inherited disease. A family may, for example, have a history of CF. By talking with a genetic counselor; family members may learn their risk of having a child with CF.

GENETIC TESTING
Tests done for clinical genetic purposes. Genetic tests may be done for diverse purposes pertaining to clinical genetics, including the diagnosis of genetic disease in children and adults; the identification of future disease risks; the prediction of drug responses; and the detection of risks of disease to future children.

H

HOME IV
Lung infections are often treated with intravenous (IV) antibiotics. Treatment can be received at home if individuals and their care providers are given enough training and support.

HEMOPHILUS INFLUZENAE (H. influenzae)
A type of *bacteria* that may colonize the lungs in CF.(Ordinary flu is caused by the influenza *virus,* not by H. influenzae *bacteria.)*

HEMOPTYSIS
Coughing up blood from the lungs. In CF this usually happens when the lining of an airway is irritated by infection (usually bronchitis). A procedure called *embolization* may be needed to plug the bleeding blood vessel.

HEP LOCK

An intravenous needle whose end can be plugged while it is not being used for medication administration.

HUFF COUGHING
A way to move mucus out of the lungs. Huffing is not the same as coughing. It is similar to the way you breathe if you want to fog up a mirror with your breath. Compared to coughing, where the back of your throat seems to close just before you release the air, huff coughing refers to when your throat doesn't seem to close. A chest physical therapist of other CF team member will to teach your child how to "huff."

HYPOXIA
A condition in which the amount of oxygen in the body is lower than normal. The ABG (arterial blood gas) test measures the oxygen level (PO_2) to tell how hypoxic a person is.

I

INFECTION
What happens when a germ (*bacteria or virus)* grows in a body tissue.

INFERTILITY
The condition of a person who can't produce children, or for whom conceiving or producing children is difficult. About 97% of males with CF are infertile because the vas deferens (the tube that carries sperm) is blocked. Many females with CF are able to conceive. Abnormal mucus may block the cervix and make it difficult for some women with CF to get pregnant.

INFLUENZA (Flu)
The respiratory illness caused by the influenza viruses. Flu is known for its sudden onset and involves headaches, muscle aches, fevers, chills, sore throat, and coughing. The particular strains of the virus that are "going around" change every year. People with CF should get flu shots every fall. The shots change each year to keep up with the changes in the virus; last year's shot may not help you fight off this year's flu.

INHALE/INHALATION/INSPIRATION
Breathing in; the flow of air into the lungs.

INTERN
A physician who has graduated from medical school and is training in a medical specialty such as pediatrics, internal medicine, family medicine, or surgery.

INTERNIST
A medical specialist in internal medicine (not to be confused with an intern).

INTRALIPID
An intravenous (IV) drip to provide extra calories, given to select patients in a hospital setting only.

IV (Intravenous)
Usually refers to an IV catheter, a small tube placed in a vein. IV antibiotics or other medicine can be given through an IV instead of, for instance, by mouth.

L

LARYNX
The part of the upper airway that contains the vocal cords. Also known as the "voice box."

LOBE
The largest division of the lung. The right lung has three lobes and the left has two.

LOWER REPIRATORY INFECTIONS
Infections involving the respiratory system below the vocal cords. They may occur in airways or lung tissue. The infections may be caused by either *bacteria* or viruses.

LUMEN
The inside of a tube. The lumen of the bronchial tubes is where the air flows.

LYMPHOCYTE
A type of white blood cell. Lymphocytes are important in fighting infection.

M

MALABSORPTION
A condition in which the body can't *absorb nutrients* normally from the intestine. When this happens, the body can't use the food that is eaten. Most people with CF do not have enough digestive or pancreatic *enzymes*, and this keeps the food they eat from being properly digested. It passes out of the body without being absorbed, causing fatty, foul-smelling stools, that leads to poor growth. Malabsorption can be controlled with pancreatic enzymes taken along with food. The enzymes help break down food in the small intestine so that it can be absorbed.

MECONIUM ILEUS
A condition of some newborn babies with CF. The intestine is blocked with a mixture of meconium and *mucus*. Meconium is a thick, black substance normally found in a baby's intestine before birth. An operation may be needed to treat the blockage.

METERED DOSE INHALER (MDI)
A medication that is delivered to the lungs with an inhaler or "puffer." Examples would include albuterol, which helps open up airways, and steroid inhalers, which control inflammation in the lungs.

MOTILITY
Movement. When used in reference to the intestines, this means the contractions of the muscles that help propel intestinal contents on their journey from the esophagus to anus.

MRSA (METHICILLIN-RESISTANT STAPHYLOCOCCUS AUREUS)
A bacteria that may colonize the lungs of patients with CF. It can be more resistant to standard therapy.

MUCOLYTIC AGENTS
Medicines sometimes prescribed to break down *mucus in the airways*.

MUCUS
A slippery, sticky liquid produced by the *mucous membranes* and mucous glands. It moistens and protects the membranes. In CF the mucus isn't normal. It may cause blockage in the lungs or intestine.

MUCOUS MEMBRANES
The tissues in the body that produce *mucus*. They line various passages of the body. For example, mucous membranes line the nose, mouth, *bronchi, esophagus,* stomach, and intestine.

N

NASAL FLARING
Widening of the nostrils with each breath (often abbreviated, "flaring"). This is a sign that someone is working harder than normal to breathe.

NEBULIZER
A cup holding liquid medicines for breathing treatments. As an air compressor forces air through the medicine in the nebulizer, the liquid becomes a mist. A person can *inhale* the mist using a mask or mouthpiece. The medicines may include *bronchodilatators* and *antibiotics.*

NEWBORN SCREENING
Tests of newborns to screen for serious treatable diseases, mostly genetic. The newborn screening tests done in the United States are decided on a state-by-state basis.

NG TUBE (Nasogastric Tube)
A tube that passes through the nose into the stomach. This tube is for feeding someone who can't eat, for continuous feeding during sleep, or for the administration of other substances such as medicines.

NUTRIENTS
Substances the body uses from *digested* food. They are needed for energy, growing, and normal functioning. Nutrients fall into several groups: proteins, carbohydrates, fats, vitamins, and minerals.

O

OXIMETRY
A test that measures how much oxygen is carried by the red blood cells. In this painless test, an instrument applied to the skin determines how well a special beam of light passes through the skin of a finger, toe or ear lobe. The test result is called *oxygen saturation* (see below).

OXYGEN SATURATION
The result of an *oximetry* (see above), measured as a percentage. It tells how much of the hemoglobin in the red blood cells is carrying oxygen. When the hemoglobin carries a full load, it's "saturated." If the test result is low, the person may need to *inhale* more oxygen.

P

PANCREAS

A *digestive* gland located behind the stomach and connected to the first part of the small intestine. The pancreas produces pancreatic enzymes, which travel into the small intestine to *digest* food. In CF, the ducts (drainage tubes) in the pancreas may be blocked by *mucus*.

PAN-RESISTANT
Resistant to all tested antibiotics.

PEAK LEVEL
The highest level of a medicine in the blood streams, as measured by a *drug level* test. Some medicines must reach certain levels to be effective. Others cause side effects when their level is too high. Peak levels are measured to determine the effectiveness and safety of some medicines. See "TROUGH."

PEP DEVICE (Positive Expiratory Pressure)
A small device used as a form of *chest physical therapy* (CPT). *Exhaling* into the device increases pressure in the airways. This may stimulate coughing. The purpose of using the PEP device is to force *mucus* up and out of the lungs. There are also PEP masks, which cover the nose and mouth rather than using a mouthpiece.

PERCUSSION
A technique for *chest physical therapy* (CPT). A cupped hand is clapped over the rib cage. This sends a gentle force through the lungs to loosen *mucus*. Percussing different areas of the chest and back helps move mucus up into the airways so that it can be coughed out.

PERIPHERAL LINE
An *IV (intravenous)* line in a small vein, usually in the arm or hand. A short catheter (tube) is inserted into the vein, taped down, and then closed off with a rubber cap. Each time the medication is given, a needle is inserted into the rubber cap. Then a syringe or pump pushes (infuses) medicine through the catheter into the vein. This is called a peripheral IV because it's placed in a vein farther from the heart than a *central line*.

PICC LINE (Peripherally Inserted Central Catheter)
A long-term catheter type that is inserted peripherally, typically the arm or hand.

PFTs (Pulmonary Function Tests)
A series of tests that shows how well a person can breathe. The tests measure the flow rate (how fast the air moves) and the volume of air moving in and out of the lungs. The tests also measure how the airways respond to inhaled medications.

POLYP
A small growth of tissue. Polyps grow out of *mucous membranes* and are usually not benign (cancerous). Some people with CF have nasal polyps. Although these are not painful, these may obstruct the passages or sinuses. A *polypectomy* may be needed to remove a nasal polyp.

POLYPECTOMY
A surgical procedure to remove one or more polyps. This is usually done by an otolayngologist (ear, nose and throat) surgeon. The polyps may reappear.

POSTURAL DRAINAGE

A technique for *chest physical therapy* (CPT). The body is placed in various positions (postures) during *percussion*. As percussions loosen *mucus*, the postures allow gravity to help drain the mucus into the large airways. Then the mucus can be cleared from the lungs by coughing.

PRODUCTIVE COUGH
A cough that brings up *sputum* or *mucus* from the lungs. This is common in persons with CF. It's also described as a "wet" or "loose" cough.

PROGNOSIS
A predication of the course or effect of a disease. It's often a way of describing the chance of recovering from a disease, or it may describe how long a person is likely to survive. Prognoses for different people with the same disease may vary greatly, and the prognosis of one person may change over time.

PRN
An abbreviation meaning "as needed" (from Latin).

PSEUDOMONAS AERUGINOSA
A common bacteria found in the lungs of CF patients.

PULMONARY EXACERBATION
A sick episode, defined by several criteria that may include: Increased cough, increased sputum production, fatigue, poor appetite, and malaise. Often treated with oral or IV antibiotics.

PULMONOLOGY
The study of the *respiratory system,* mainly the lungs. Doctors who specialize in this area are pulmonologists.

R

RECTAL PROLAPSE
The inner lining of the rectum comes out through the anus. In rare cases, surgery may be needed. Rectal prolapse may be the first sign of CF in young children.

REFLUX
The backward movement of fluid; this term is often used to refer to gastroesophageal reflux.

RESEARCH
In **basic research**, scientists seek to increase our knowledge of basic processes. They might try, for example, to understand chemical processes in certain cells, then see how these processes are different in CF. Researchers might also look how these different processes cause the problems of CF.

In **applied research**, scientists take the findings of basic research and use them to solve problems of everyday life. They might, for example, look for ways to change the CF *gene* to reduce or negate the problems in CF.

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

RESIDENT
A physician who has completed medical school and is undertaking further training in a medical facility.

RESISTANCE
Sometimes bacteria become resistant to an *antibiotic*. This means that antibiotic can no longer control or kill that kind of bacteria.

RESPIRATION
The process of breathing.

RESPRATORY RATE
The number of breaths taken in one minute.

RESPIRATORY SYSTEM
The parts of the body used for breathing and respiration. The upper respiratory system includes the mouth, nose, and throat. In the lower respiratory system are the trachea, *bronchi, bronchioles, alveoli,* and lung tissue.

RETRACTING
The pulling of skin between the ribs with each breath, indicating hard breathing.

RSV (Respiratory Syncytial Virus)
The most common cause of viral respiratory *infections* in children younger than five. RSV can cause colds, *bronchitis,* and *pneumonia.*

S

SALINE
A mixture of salt and water at a level similar to the body's own fluids. Saline is mixed with antibiotics when they are given IV. It's also commonly used to dilute medications for breathing treatments.

SEDATION
A state of reduced excitement, anxiety, and (often) a state of mildly reduced consciousness. Many drugs are used to produce sedation and are commonly used to decrease anxiety during medical procedures.

SENSITIVE
This term means "killed by" when used to describe bacteria's relation to an antibiotic. For example, the statement, "*Streptococcus* bacteria are sensitive to penicillin," means that, in the laboratory, penicillin kills *Streptococcus* organisms.

SENSITIVITY
A laboratory test. The test is administered after *bacteria* have been identified in a *culture* from a patient. (In CF, this is usually a *sputum* sample.) The bacteria are exposed to different kinds of *antibiotics*. The results show which antibiotics work best in the lab to kill or control the bacteria. The bacteria are said to be sensitive, or killed by the antibiotics. This test helps doctors choose the best treatment for a patient's infection.

SIGNS

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

SINUSITIS
An inflammation or infection of the sinuses; the airspaces within the bones of the face.

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

STAPHYLOCOCCUS AUREUS (S. aureus)
A common bacteria found in the lungs of CF patients.

STERILITY
The inability to produce children. In women it means being unable to conceive (become pregnant). In men it means being unable to cause conception (pregnancy).

STOOL ELASTASE
A diagnostic test to determine if a person with CF has pancreatic function.

SWEAT TEST
A method of diagnosing CF. The sweat glands are *exocrine glands,* and with CF they produce sweat that is saltier than normal. CF is diagnosed when the chloride level in the sweat is elevated. The test is painless, relatively quick, and is the gold standard for CF testing.

SYMPTOMS
The things people notice when they are ill or have other problems. A person with CF may feel tired or have chest pain with an infection. Stomach cramps may be a symptom of malabsorption. A *sign* of an illness, in contrast, is something physical that can be observed by another person, such as a fever or *crackles*.

T

Total Parenteral Nutrition (TPN)
With TPN, the body is given *nutrients through* a tube (*central catheter*) into a large vein. This lets the body receive and absorb the nutrients directly from the bloodstream, without food going through the *digestive system*. The solution used typically contains dextrose (a form of sugar) amino acids (the building blocks of protein) minerals, electrolytes, vitamins, and lipid (fat).

TRACHEA
The windpipe that connects a person's lungs to the nose and mouth.

TROUGH
The lowest level that a drug reaches in the bloodstream (this level is found immediately preceding a dose of the drug). See "PEAK."

TOXICITY
Harmful effect(s). This term is often used to refer to the undesirable effects of a medication.

U

UPPER RESPIRATORY INFECTIONS (URI)
Infections involving the *respiratory system* above the vocal cords, including the nasal passages, sinuses and pharynx (throat). The infections may be caused by *viruses* or *bacteria*. The common cold, for example, is a URI that is caused by a virus.

V

VACCINE
A preparation of a weakened or killed pathogen, such as a bacterium or virus, or of a portion of the pathogen's structure that upon administration stimulates antibody production or cellular immunity against the pathogen but is incapable of causing severe infection.

VACCINATION
Administration of a *vaccine* to a patient. The purpose is to help the body build *resistance* to a specific disease.

VIBRATION
A method to help loosen and remove mucus from the lungs. It's a type of *chest physical therapy*.

VIRUSES
Organisms smaller than *bacteria*. Viruses cause certain *infections* and diseases that can't be treated with the usual *antibiotics*. Only a few viruses have specific medications to treat them. Viral illnesses (those caused by a virus) include chickenpox, mumps, measles, the common cold, and *influenza* (flu).

W

WHEEZING
A high-pitched whistling sound heard in the lungs. It may be heard with a stethoscope or, at times, with normal listening. It happens when air moves through narrowed airways. It may be a sign of asthma.

SUPPLEMENTAL INFO