Seattle Children’s offers interpreter services for Deaf, hard of hearing or non-English speaking patients, family members and legal representatives free of charge. Seattle Children’s will make this information available in alternate formats upon request. Call the Family Resource Center at 206-987-2201.

This handbook has been reviewed by clinical staff at Seattle Children’s. However, your child’s needs are unique. Before you act or rely upon this information, please talk with your child’s healthcare provider.

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### Personal stories/testimonies

**Cystic Fibrosis: the Ultimate Teen Guide (It Happened to Me series, #14)**


**The Power Of Two: A Twin Triumph Over Cystic Fibrosis**

( good for young adults)

2007. Byrnes, Anabel and Isabel. Twin girls tell of their lifelong struggle to pursue normal lives with cystic fibrosis while grappling with the realization that they will die young. An honest and gripping portrayal of day-to-day healthcare, and the impact of chronic disease on marriage and family.

**The Spirit Of Lo: An Ordinary Family’s Extraordinary Journey**

2000. Dietrich, Don and Terry. An ordinary family is faced with an extraordinary challenge — a child with cystic fibrosis. This family’s story covers topics like grief, infections, insurance concerns, drugs, and all the issues families have. What emerged is The Spirit of Lo which enables the family and their community to face each new day of life’s dance on the edge of mortality.

### Stories for children

**Miguel And Sarah: Close Friends And Cystic Fibrosis**

( good for ages 9 to 12)

1999. Dowell, Andrea. Miguel explains what he has learned about Cystic Fibrosis from his friend, Sarah, who has the disease.

**Taking Cystic Fibrosis to School**

( good for ages 4 to 8)

2000. Henry, Cynthia. Jessie explains to her classmates her condition, cystic fibrosis. Even though she has cystic fibrosis, she can still attend school and do many of the same things as her classmates.

### Inspiration

“Welcome to Holland” — copyrighted Emily Perl Kingsley poem [http://users.erols.com/jmatts/welcome%20to%20holland.html](http://users.erols.com/jmatts/welcome%20to%20holland.html)

### Transition to adult care

University of Washington Adult CF Clinic

206-598-8446
Resources

Websites
- Cystic Fibrosis Foundation
  www.cff.org
- Online community
  www.cfvoice.com
- Resources for school teachers
  http://www.cff.org/LivingWithCF/AtSchool/
- CF Legal Information Hotline
  CFLegal@cff.org
- Healthy Web surfing
  www.cff.org

Books

Informational

An Introduction to Cystic Fibrosis: For Patients & Families
2003. Cunningham, James M.D. and CF Foundation. Guide for patients and families. Provides definition of CF, how it affects the various parts of the body, and living with CF.

Cystic Fibrosis: A Guide For Patient And Family
2004. Orenstein, David. Offers practical information on day to day concerns such as school, travel, exercise, nutrition, medications, physiological effects, long term issues and prospects of a cure. Provides answers to frequently asked questions by patients and families. Third edition.

Parenting Children with Health Issues
2007. Cline, Foster, MD.; Greene, Lisa. Principles of parenting a child with special health issues. Also covers growth and development, siblings, emotional health and mental health topics.

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**Work and career:** When considering a job or career, your young adult child should think about their health. Some people with CF do best in careers that are not too physically demanding (like professional athletes). Your child should also consider jobs or careers that won’t expose them to cigarette smoke, dust, chemical fumes, or people prone to infections (like healthcare positions or work with small children or the elderly).

When a person interviews for jobs or careers, they do not have to tell employers that they have CF. It is illegal to ask if you have a disability, but they can ask you if you have the skills and are physically able to do the job. Some people with CF choose to tell their employers from the very beginning so they can ensure that their schedule can be flexible, part-time, or includes an adequate amount of sick and vacation time to help support them if they need to be hospitalized or to stay home sick. Ultimately it is up to your child if they tell their employer about their disease.

**Lung transplant:** One treatment for CF that people may start to consider in their teen and young adult years is lung transplantation. Lung transplantation is a treatment and not a cure. The new lungs will be disease-free, but the rest of the body still has CF. Lung transplants are expensive and have many risks. This decision should be discussed with your family and doctor at length.

Lung transplantation is done at the University of Washington Medical Center (UWMC). After you have a lung transplant, UWMC will follow up on all your CF care regardless of your age.

**Complete transition to adult care:** The University of Washington Medical Center (UWMC) has a clinic for adult cystic fibrosis patients. We encourage our patients to transition to this clinic when they are around 18–21 years old, but patients can continue to come to Children’s Hospital until the age of 21 years.

By your child’s 21st birthday, their CF care should be fully transferred over to UWMC. The CF team will help your family with this process when your child is in their teen years. We help make sure the transition goes as smoothly as possible. The CF care team will miss your family as your child “graduates” into the adult world of CF care. But, we will be happy to see a fully grown, happy, healthy person living an adult life.
Getting started

Using this handbook

Welcome to the Cystic Fibrosis Clinic at Seattle Children's Hospital.

This handbook is a tool to use in your journey after a diagnosis with cystic fibrosis (CF). You are likely filled with questions about the disease and the future of your child's life. This handbook will help you get a better understanding of CF and its treatment, learn skills to help clear your child’s lungs and see how the CF Clinic fits into your life. Keep this handbook as a reference over the coming years.

Though getting a diagnosis of CF is hard for a parent, know that things will be OK. You have every reason to feel positive about your child’s future.

We will partner with you on your journey.

How to contact the CF care team

If you have a question:

- During the weekdays — call the CF clinic coordinator/program assistant at 206-987-2024.
- At night or over the weekends, for urgent issues — call 206-987-2000 and ask for the "pulmonary doctor on call."

Your primary care provider (your pediatrician or family doctor) is an important part of the team and can often be your first contact. Having CF is only one part of your child's total health and development. The CF Team will work with you and your primary care provider all along the way.

See page 33 for more information on who to call, when.

Phone numbers for key CF Team Members

- Cystic Fibrosis Nurses
  206-987-3316
- Dietitian
  206-987-1744
- Respiratory Therapists
  206-987-2258
- Scheduling (Family Service Coordinator)
  206-987-5610
- Social Worker
  206-987-4144
- Clinic Coordinator/Program Assistant
  206-987-2024

Young adulthood

Things to consider as a young adult

The issues outlined in adolescence continue into adulthood. At this stage, many people with CF become completely independent in the care of their disease as they go off to college, move out of their parent’s house, start careers, and get married.

Dating, marriage, and having a baby: The same rules of dating apply to people with CF as apply to people without CF. Nobody should be pressured into getting intimate if they are not ready, and when they are ready for intimacy, they should take precautions to prevent STDs by using barrier methods like condoms. Keep in mind that it is possible for women with CF to get pregnant, and on the rare occasion men with CF can get women pregnant, so precautions should be taken to prevent pregnancy until both partners are ready.

People with CF can have a very healthy and normal sex life. Planning to have a family when one person has a chronic illness has many difficult challenges and this decision should also be discussed with your CF doctor. Both partners need to consider how pregnancy will affect the body of the female who has CF. Infants born to mothers with CF will carry the CF gene or could also have CF if the biological father carries the CF gene.

Education: Many people with CF complete further education after high school, graduate from college, and even go on to attain advanced degrees. The decision of where to go to college can be challenging and even more difficult for someone with CF. They must make a decision about attending a college near home, close to friends and family, or attending college away from home where they may not have the resources they are used to in order to help care for themselves.

If they do choose to attend college away from home, they will mainly need to make sure that they can continue their airway clearance independently with the Flutter, Acapella vest, or whatever ACT works best for them.

If they attend a college away from the family, it may be tempting to try and leave CF behind and neglect self-care. It is crucial that your young adult child continues their daily treatment regimen in order to keep their lungs healthy and prevent any decline. It is also important to keep healthy by eating right and continuing physical activity while off at college as well.

Things to consider as a young adult

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Transitioning into adult care: Seattle Children's Hospital treats children from infancy up to the age of 21 years. After the age of 21 years, adults with CF continue their care at the University of Washington Medical Center (UWMC). Therefore, during the teenage years, the CF team will start to help you and your teen through the transition process of CF care at Children's to UWMC. This can be an anxious time for teens and families who are used to getting care from Children's CF team for most of their life. However, it can also be a very exciting time, because it means your teen is often transitioning into other adult aspects of their life too such as college, their own apartment, or a job. The CF team at Children's will work very hard to make sure that you have a smooth transition into adult care at UWMC. Please feel free to ask us questions or tell us how we can make this transition go well for you and your teen.
What is cystic fibrosis?

Children with cystic fibrosis have glands throughout their body that produce thicker than normal secretions (mucus). These thick secretions block the normal flow of mucus in many organs. The lungs and the pancreas are two of the most important organs affected.

The lungs

When mucus clogs the lungs it can make breathing difficult. The thick mucus clogs the air tubes and can cause germs to grow in the airways. These germs cause inflammation or swelling and infections that can lead to lung damage. To help reduce the infections, we use antibiotics, airway clearance techniques (removing mucus from lungs) and inhaled medicines. Despite all these treatments, over time, permanent lung damage does occur.

The pancreas

The pancreas is a very important organ that produces enzymes used to digest food. In most but not all children with CF, the pancreas is damaged or clogged with mucus. It cannot send enough enzymes to the small intestine to digest food. As a result, your child has large, bulky bowel movements. They also lose nutrients from their food that could not be absorbed. Your child will need to take enzymes in the form of pills. The enzyme pills allow your child to digest food properly.

Hospitalization: The teenage years may be the first time your child needs to be hospitalized for a CF exacerbation. At this point, your child is central to their own care. The CF team will explain what to expect, how to participate in their own care, and what choices they can make about their own care. It is hard for teens to be in the hospital because they are away from their friends, school, and regular activities. Seattle Children’s encourages teens to stay in contact with their friends via phone, Internet, and by friends visiting the hospital. Children’s also has a special Teen Zone that parents and healthcare staff are not allowed to enter. It has video games and is often the venue for fun make-up parties or movie nights.
What cystic fibrosis is NOT!

- **Cystic fibrosis is NOT asthma.**
  Although some doctors misdiagnose CF as asthma, they are two totally different conditions with some similar symptoms like wheezing, tight chest and cough.

- **Cystic fibrosis is NOT contagious.**
  CF is a hereditary disease (passed on from your genes) — that cannot be passed from person to person by any form of personal contact like coughing or touching. It can be passed genetically, should your child have a baby.

- **Cystic fibrosis does NOT make you look different.**
  Children with CF look the same as other children. There are no physical outward signs of CF.

- **Cystic fibrosis does NOT affect brain development.**
  Children with CF learn and develop on schedule, within normal limits or beyond.

- **Cystic fibrosis is NOT a disability.**
  While some children with CF have some limitations due to their disease, most are able to lead normal, active, and productive lives.

Many women with CF can get pregnant. If your teen chooses to be sexually active it will still be important for her to use some form of contraception and protection from STDs. In addition, your teenage girl can still get STDs and should therefore use condoms to try and reduce her risk of exposure. Vaginal yeast infections may occur due to the side effect of taking antibiotics for lung infections. This should be reported to your daughter’s doctor so she can get treatment.

**Nutrition:** Teens will need to continue to take pancreatic enzymes before each meal and require a high-calorie/high-protein diet. Nutritional needs are high during adolescence. This is a time of life when caloric needs are very high. In general their diet should not be restricted. Continue to work with the school regarding medicine use on campus. Children with CF do have a risk of developing a CF-related form of diabetes. Diabetes means that your body has difficulty moving glucose (sugar) from your blood into your cells so that your body can use the glucose for energy. Since about 10–15% of teens with CF develop diabetes, your doctor may recommend a glucose test during adolescence.

**Physical activity:** Just as it is for a small child, exercise is important for your teen to stay healthy. It can help clear their airways of mucus, help them feel good about themselves, and give them a forum for social activity. Teens with CF can still do the same physical activities that other teens do such as cheerleading, running track, dance, basketball, swimming and lifting weights. To see some stories about teens with CF involved in sports visit www.cfvoice.com and watch the teen videos on the “Ages 13-17” page.

The same advice should be used for teens engaging in physical activity as for school-age children. Teens should be reminded that if they feel winded or fatigued during exercise they should slow down and take a short break. They should not “overdo it” regardless of which physical activity they choose.

**Independence and self-care:** During the teen years your child is at an awkward place where they still rely on you for some parts of their care and yet they are becoming increasingly independent in other areas of their care. During the adolescent years it is important for your child to become more involved in their care.
Basic genetics of CF — how did my child get cystic fibrosis?

About 30,000 children and adults in the United States have cystic fibrosis (CF). It is a genetic disease and is not contagious. Genes lie within your DNA and control the traits and functions of our body, like cell functions, hair and eye color.

You can’t catch CF from someone. Cystic fibrosis is caused by mutations (abnormalities) in one gene within your DNA. This gene codes for a protein called CFTR (Cystic Fibrosis Transmembrane Conductance Regulator). This protein is needed to help salt move across cell walls. A child with CF has too little of the CFTR protein. This results in thick mucus and salt loss from the body.

Every person has two copies of the CFTR gene, one inherited from the mother and one inherited from the father. People with CF have a mutation in both copies of this gene. Their mother and father are almost always carriers with one working copy of the CFTR gene and one copy with a mutation. Carriers do not have CF themselves. Every pregnancy between two CF carrier parents has a 25% (¼) chance to have CF, a 50% (½) chance to be an unaffected carrier, and a 25% (¼) chance to be an unaffected non-carrier.

See the diagram below. For more information on genetics, see the appendix.
Signs and symptoms of CF

The most common signs and symptoms are listed below, although they vary some from person to person:

- Very salty-tasting skin
- A large appetite, but poor weight gain
- Greasy, bulky bowel movements
- Chronic coughing or wheezing
- Many lung infections, like pneumonia and bronchitis
- Frequent sinus infections

Sometimes children are told that they have asthma or chronic bronchitis when they really have cystic fibrosis. New research shows that the severity of CF symptoms is partly based on the types of CF gene mutations (defects). Scientists have found more than 1,500 different mutations of the CF gene.

How is CF diagnosed?

Most children are diagnosed with CF at birth or before the age of 3. Washington state began screening newborns for cystic fibrosis in March of 2006. Infants with a positive newborn screen will be ordered a sweat test. For older children, a doctor who sees the symptoms will order either a sweat test or a genetic test to confirm the diagnosis.

A sweat test is the most common test used to diagnose cystic fibrosis. Two small electrodes are placed on the skin (usually on the arm) to stimulate the sweat glands. Sweat is then collected and the amount of chloride, a part of salt in the sweat, is measured. See photos, left.

In a genetic test, a blood sample or a sample of cells from the inside of the cheek is taken. It is sent to a lab to see if any mutations of the CF gene are found. A genetic test can be used if the results from a sweat test are unclear.

What about school work while my child is in the hospital? It will be important to work with your child’s teachers to obtain homework while your child is hospitalized. If you need help contacting your child’s school or teachers our CF team would be happy to help. At Children’s Hospital, we also have a school room for your child to study in if they choose. However, many children with CF choose to complete homework in their patient rooms and are easily able to keep up with their peers at school.
Living with cystic fibrosis — a typical day

Children who have cystic fibrosis, just like any child, can be bright, enthusiastic, and may often be precocious. Most children with CF are healthy for many years and can take part in all family activities. Of course, there will be times when your child’s chronic illness will put stress on your family. You may feel you are on a roller coaster of ups and downs with your child’s care in the beginning. Take care of yourself, connect with others and embrace the help and support of others.

Need for ongoing well-child care

Children with cystic fibrosis should have a primary healthcare provider near their home. Your child will need to be seen for routine immunizations, regular childhood illnesses, and well-child care. The CF Center staff will work closely with your child’s regular healthcare provider and coordinate care as needed.

In most cases, CF patients are seen in the CF Clinic every three months in order to help them stay healthy and to catch any signs of illness early.

What is a typical day for someone with CF?

Each day most people with CF:

- **Take pancreatic enzyme supplement** capsules with every meal and most snacks (even babies who are breastfeeding may need to take enzymes).
- **Take multivitamins**.
- **Take aerosolized medicines**. These are liquid medicines that are made into a mist or aerosol and then inhaled through a nebulizer or metered dose inhaler “puffer.”
- **Do some form of airway clearance or chest physical therapy**, at least once and sometimes up to four or more times a day.

Need for a Smoke-free Home

Breathing in someone else’s cigarette smoke is not a good idea for anyone, but it’s especially harmful for children with CF.

Your child’s lungs are more susceptible to damage.

Remember it’s up to the adults to protect the children from secondhand smoke.

Please don’t allow smoking in your home or car, and make sure your child never starts smoking.
Meet Ellie

We found out our daughter, Ellie, would have CF when I was 5 months pregnant. I have a cousin with CF and another who passed away from the disease so I was aware of what this diagnosis meant. We were devastated, scared, and angry and felt totally helpless all at the same time. Before the birth we were able to meet with the CF team at Seattle Children's Hospital. They spent time with us answering our questions, dispelling our fears and updating us on the current advancements. We left that appointment feeling like we were not alone in this and hopeful for our daughter's future.

The first few months following her birth were hard. It was not only challenging having a newborn, but then we had enzymes, vitamins and treatments to get used to as well. We felt very protective and wanted to isolate ourselves in an attempt to keep her as healthy as possible. I had fears that she would never be able to do the normal childhood activities. The regular appointments at Children's were extremely helpful. We had so many questions and concerns. It was reassuring to know we were in good hands and the support given by the team was invaluable. We always felt a little more grounded and less fearful when we left the clinic.

Now Ellie is 5 years old and will be starting kindergarten in the fall. She is living a healthy, normal life. She has never had a lung infection nor has she had to be hospitalized. The doctors are on top of things before they become a problem. We visit the CF center for regular check-ups every 3 months and make sure she gets her medications and treatments daily. We are hopeful for a cure, but until then we are focusing on the constant advancements being made to improve the quality of life in those with CF.

— Laura and Don

School-age child

CF at school

Children with cystic fibrosis tend to do very well in school, even when they are absent for illness, but problems may arise due to these issues:

- Embarrassment caused by frequent use of the bathroom and smelly bowel movements
- Embarrassment about coughing in class or taking enzymes in the school lunchroom
- Difficulty keeping up in recess or gym classes

Meet with the school staff at the start of the year. This can help you and your child avoid most of these difficulties. If you feel uneasy about this task, the clinic staff would be happy to help. The Cystic Fibrosis Foundation also has a very helpful website for teachers and parents about CF issues at school:
http://www.cff.org/LivingWithCF/AtSchool/

You can print off materials from this Web site for your child's teacher or direct the teacher to visit this Web site for more information. The school nurse in your child's school district can also be helpful in coordinating your child's care (e.g. medications, ability to have breaks from class, etc.) while at school and help educate teachers about CF.

Nutrition: Your child will need to take their pancreatic enzymes before every meal and snack, even at school. Become familiar with your school district’s policy on medicines that children can carry with them at school.

Children with CF need to eat a diet high in calories in order to support their growth. Parents can help support their child’s growth by offering frequent high-calorie and nutritious snacks. Ask your dietician for specific ideas for your child.

Beginning independence and self-care: As children with CF grow up, they start taking on more responsibility for their self-care. Just as they learn to cook simple meals and do chores around the house during the school-age years, your child should also start to learn how to care for their CF. During school-age years parents still have most of the responsibility for their child’s medicines and treatments. But, now is a good time to gradually start teaching your child how to take care of themselves. If parents can help their child to grow toward complete self-care
Infants have very tiny air passages, and colds are more likely to develop into lung infections or pulmonary exacerbations. Some parents may choose to delay sending their infant to daycare for the first few months of life. However it is impossible to prevent all colds in your child. Your toddler and preschool-age child should learn hand washing, just like other children, in order to help prevent illnesses. A smaller daycare setting with fewer children may somewhat decrease the amount of colds your child is exposed to.

**Hospitalization:** Infants with CF, on a rare occasion, need to be hospitalized for short periods of time. This is usually for pulmonary exacerbations — when an infection results in breathing trouble. If your infant needs to be hospitalized, the best thing you can do as a parent to comfort your baby is to be by their side and to keep familiar routines. The hospital can be scary to infants and toddlers because it is an unfamiliar environment. Having a parent present to cuddle and rock them or to have a favorite toy or blanket with them often helps the infant to cope.

Toddlers and preschool-age children are also rarely hospitalized. At this age children are able to understand a little more about the hospital. They are mostly afraid of being abandoned or are unhappy about not being able to play freely because of the IV lines. Children in the hospital still need to play, and many forms of play can be done while they have an IV. As long as they wear a mask outside of their hospital room to protect them from illness, they may go to the hospital playroom. You may also choose to have your child stay in their hospital room and engage in play, just as they would at home in their own bedroom or play room.

The hospital has all sorts of unfamiliar sights, sounds, and smells. Again, your presence with your child in the hospital can help quiet their fears. Because of their developmental stage, preschool-age children may also view hospitalization as a punishment for something they imagine they did “bad.” Toddlers and preschool-age children are also learning to assert their independence and may try to refuse treatments. This is normal, and the best way to help the child through medications and procedures is to give them simple explanations and to assist the healthcare worker to complete the task as quickly and smoothly as possible.

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**Meet Brian**

My son is an active 20-year-old who goes to college and loves to hang out with friends and play tennis and video games — and, oh yes — he has cystic fibrosis.

Brian was diagnosed as an infant, so CF has always been an accepted part of life. When he was younger, time for inhaled medications and airway clearance (which we call PT) was always family time — spent cuddling or playing games. (Brian can play a video racing game in any position and still beat everyone in the family!) As he got older and had friends around, card games and imagination games continued uninterrupted. During the early years of school, PT was also a great time to review spelling words, or math facts, and it’s still a good time for studies — or a break from studying.

Trips to clinic have often been family time as well. Clinic visits in the morning with the CF team are usually full of smiles and laughter (it’s the best medicine, you know), followed by trips to the zoo or science center or some other adventure. It’s a great excuse to take time out to have fun!

CF does cause stress at times — those blood draws, that first tune-up (going into the hospital for IV antibiotics) — but it has made us a close-knit family that appreciates every day, and takes the time to enjoy each other.

Things have changed quite a bit now that Brian’s in college. He’s making new friends, trying new things (his latest passion is jujitsu) — and is working hard towards a career. He has a great internship working on computer programs for an environmental consulting company, and is continuing his college studies. He is planning a long, successful career and a happy life where CF care is a necessary part, but not the central part.

Brian will be leaving the Children’s CF clinic in a few months to go to the adult CF clinic. Yes, CF makes for a bumpy road at times, but we’ve had a great team — the clinic staff at Seattle Children’s — to ensure that Brian gets to this point in the best possible shape, with hope for the future and for a cure.

So as you travel this path, know that many others are doing it with you, with great joy and success, and that you’ve got wonderful partners at Seattle Children’s Hospital who are with you every step. We are so thankful for our team. We couldn’t have done it without them.
Your baby’s first years:
Infancy through preschool age

Diagnosis as a newborn or infant
If you were living in the state of Washington at the time of your infant’s diagnosis of cystic fibrosis (CF), likely you found out by newborn screening. Although this is sometimes a sad and confusing time for parents and caregivers, healthcare professionals are confident that early diagnosis of CF allows for a healthier child. The earlier we start preventive care, the better.

Daily life in infancy through preschool
Developmental milestones: Your baby will be just like other babies at this stage. They will eventually learn to sleep through the night, occasionally get colds and the stomach flu, crawl, learn to walk, and become potty-trained. Toddlers will cry and may have temper tantrums. In the child with CF, crying can lead to a coughing attack. This coughing may frighten you and other adults, but keep in mind it is not harmful. Coughing helps bring up mucus.

Nutrition: Infants with CF can breastfeed or drink formula for the most part. Sometimes special formulas are needed. You may need to give your infant pancreatic enzymes right from the start. Children from 1 to 3 years of age begin to take control of what foods they like and begin testing limits. Many toddlers go through periods of refusing certain foods. It is not unusual for parents with toddlers and preschoolers to be frustrated with a child’s control issues at mealtime. The CF dietitian can help you with this and with planning meals and snacks.

Potty training: Children with CF will gain control of their bladders during their toddler and preschool-age years just like children without CF. Bowel training (going poop in the toilet) sometimes occurs later in children who have CF. This is because of large and frequent bowel movements. Do not be too concerned if your child is not fully toilet-trained until age 3 or 4 years.

Daycare: Many parents or caregivers use daycare. Children in daycares often get exposed to more colds than do children who stay at home. However, children with CF get the same number of colds as other children who attend daycare. No more, no less.
Your child’s nutrition

Calories in the diet
Children with CF may have a hard time gaining weight. This is because their digestive enzymes don’t reach the intestines. Without these enzymes, fats and proteins cannot be properly absorbed. Carbs aren’t affected much. Also, infections can cause children to have a hard time gaining weight.

Your child needs about 30–50% more calories and twice as much protein as other children throughout their life. Eating this high-protein, high-fat and high-calorie diet will help your child stay in the recommended 50th percentile or above body mass index (BMI) range. The best meal plans include frequent meals and snacks! Nutritious, high-calorie snacks play a key role in providing enough calories.

The CF dietitian will help you with your child’s nutrition. She can tell you if your child is getting enough nutrition by tracking weight gain and growth, by measuring body fat and muscle stores, and by checking the total daily calories. To find out if your child is getting enough calories and protein, you may be asked to keep a three-day food record. These food records are computer analyzed for nutrient composition and then compared to your child’s requirements.

Sometimes, high-calorie liquid supplements are needed. These can be taken by mouth or through a feeding tube. The dietitian may suggest a special formula or high-calorie dietary supplement for your child. See the section on supplements.

Your child’s development across the years
Throughout childhood, typical problems will come with growing up. Having an illness such as cystic fibrosis can add to these “growing pains” and may create more stress on your family. The next few pages talk about some common concerns at milestones of your child’s life.

Many concerns don’t have definite answers. We hope you will discuss your concerns with the clinic staff or other parents. Parents of an older child with cystic fibrosis can often be a good source of support and information.

The Family Resource Center at Seattle Children’s also has resources for you. Call 206-987-2201 for books and handouts on growth and development, parenting and living with a chronic illness.
A review of the basic food groups

Each day, your child will need:

**Carbohydrates**
These foods (sugars, starches, fiber) provide quick energy and calories to help in growth. They are the easiest to digest. Examples of carbohydrates include breads, cereal, pasta, fruits, vegetables, sugar, and pastries.

**Protein**
Protein is needed to build and repair the body as well as help fight infection. Children with cystic fibrosis should eat more protein than other children. Foods high in protein include meats, fish, eggs, cheese, milk, nuts, beans and yogurt.

**Fat**
Fat is the most concentrated form of calories and is used as the body’s energy source. Like proteins, fats may be poorly absorbed by children who have CF. Fats called essential fatty acids (EFAs) are important nutrients in the diet. Foods high in EFAs include safflower, corn, sunflower, soy, and cottonseed oils.

**Vitamins and minerals**

Vitamins A, D, E and K are absorbed into the body with fat. People who have trouble digesting and absorbing fats may not always absorb enough of these vitamins. Pancreatic enzymes will help absorb the vitamins, but many children with CF need to take a special multivitamin pill every day. This is a more easily absorbed form of vitamins A, D, E and K. Vitamin A is needed for eyesight and healthy skin. Vitamin D is necessary for absorption of calcium and bone health. Vitamin D is found in milk. Exposure to 10 minutes of sunshine per day without sunscreen can also help you get more vitamin D. Vitamin E is needed for healthy red blood cells and vitamin K is essential to the clotting of the blood.

We will measure the level of these vitamins in the blood each year to tell you if your child is absorbing enough. The CF clinic will prescribe the amount of vitamins your child should take. Vitamins E and A labeled “water-miscible” on the bottle are better absorbed and should be used instead of “fat-soluble” brands. You may be given a prescription for a multivitamin with water-miscible forms of A, D, E, and K. Too much of any

Your other children

Care of your child with cystic fibrosis may be time-consuming and it may not be possible to give equal time to each child. Set time aside for uninterrupted activities with each of your children. Brothers and sisters will always fight or argue as a part of the growing-up process. It is tempting for parents to side with the child with the chronic illness. This preferential treatment can make the brother or sister feel guilty, or resentful. Try not to take sides. If fighting becomes a real problem, it may be helpful to sit down and talk with your well child alone about his feelings towards the child with CF.

All members of the family are welcome in the CF Clinic at Children’s. We would like to meet and talk with brothers and sisters. Please don’t wait until problems arise to include them in the visits.
How do I talk to others about cystic fibrosis?

What is the best way to talk with others about CF? Here are some guidelines to follow when you are asked questions.

Telling your child

When talking to your child about CF, two things to keep in mind are the age of your child and the degree of lung involvement. Do not think that you need to answer all of your child’s questions at one time. Be open and honest and ask us if you need help deciding what should be shared.

The very small child will usually remember the name “cystic fibrosis,” that it affects the stomach and lungs, and requires daily medicines. For school-age and teenage children, ask your child what they already know, correct any wrong information, and do not push beyond their immediate concerns. If your child is asking questions that make you uncomfortable, your doctor or the clinic staff can help you.

Telling others

Each circumstance is different, and will dictate the amount of information you want to share. At least give the name of the disease and a short definition of what is involved; like, “Cystic Fibrosis is a genetic disease of the glands inside the body. They make too much sticky thick mucus.”

- Neighbors may need to know that the cough is not contagious to their children.
- Teachers may need to know that your child may have to use the bathroom frequently (because of stool issues) and may cough in class.
- A camp counselor may need to know that your child has to take medicine (enzymes) with meals.

When your child enters a new situation such as school, camp, or daycare, have a short meeting with the staff in advance. The nurses and social worker in the CF clinic are happy to talk with school teachers, daycare providers, and any others who have concerns.

vitamin can be harmful, so be sure to give only the dosages recommended by your doctor. Lastly, bone health depends on adequate calcium intake and absorption. Milk and dairy products are the best sources of dietary calcium for people with CF.
Taking pancreatic enzymes

Most children with cystic fibrosis need to take pancreatic enzymes to help the intestines absorb food. A fecal elastase test is done to find if your child makes pancreatic enzymes on their own. Symptoms that the pancreas is not producing enzymes on its own include greasy stools and/or poor growth even though eating a lot of food.

Enzymes are capsules containing lipase, protease, and amylase to help people with CF absorb their food. Your child will need to take enzymes right before each meal and snack throughout life. There are many brands and strengths of enzymes.

How many enzymes does my child need?

Every child with CF needs a different amount of enzymes. Some reasons for these differences are:

- the amount of fat in the diet
- the amount of acid in the stomach
- the type of enzyme

Your CF doctor and the dietitian will help you and your child adjust the enzymes to achieve the best digestion possible. Underdosing and overdosing of enzymes can have side effects, therefore any change in dosage needs to be discussed with the CF staff.

Your child's growth is the best measurement of absorption. Regular check-ups, which include careful growth measurements, will be needed as your child ages.

Breastfeeding and formulas

Breast milk is recommended as the sole source of food for your baby for their first 4 to 6 months of life. You will need to give your baby enzymes along with your breast milk.

You may choose to give standard infant formulas like Similac or Enfamil, along with enzymes. Sometimes more specialized formulas are recommended.

Encouraging. Active research is ongoing and both scientists and doctors are hopeful that treatments to improve survival and quality of life for CF patients will become available in the near future. Also, research is ongoing to develop medicines to treat the infections and inflammation that are common in CF.

In 1998, the National Cystic Fibrosis Foundation started the Therapeutics Development Network to further research efforts in our nation. This network consists of Centers across the country, which work together to conduct the highest quality, state-of-the-art research. The coordinating center, which organizes all of the centers, and one of the research centers, are here in Seattle. It is hoped that a coordinated effort by researchers from across the country will lead to new therapies for both treatment and a cure. Several active research studies are ongoing here at Children's, and at some point, you may be given the option to join a study.
Is there a cure for CF?
A look at the research

There is no cure to date, but we are working hard to find one. While in 1960 few children with CF reached school age, now many are living past age 40. Good medical care and early diagnosis can make a difference in your child’s life span.

The research
The 1980s and 1990s brought major advances in cystic fibrosis research. Scientists discovered that the thickening of mucus is most likely due to a loss of salt and water (dehydration) from the mucus. Normally, the salt, water, and mucus are secreted by the cells that line the airways (bronchi) of the lungs. The same type of cells (epithelial cells) also line the bile ducts in the liver, the pancreatic ducts, and the intestinal tract. In people with CF, these lining cells cannot secrete salt in a normal fashion because one chemical in salt (chloride) does not pass through the cell wall as it should.

At first, scientists thought that the channels (holes) in the cell membrane, which allow passage of chloride, were missing. Now, they know that these channels are present, but a protein (CFTR) that acts as a gate to regulate flow of the chloride molecules is abnormal in people with CF.

In 1989, a group of scientists discovered the gene that makes the CFTR protein. In 1990, scientists located this regulator protein. They are still trying to understand more about how it works. The name of this protein is the Cystic Fibrosis Transmembrane Conductance Regulator protein, or CFTR. Scientists hope to find medicines in the near future that will allow this protein to work normally. Then, chloride movement in cells will no longer be abnormal.

In 1990, scientists took a copy of this gene from a person who did not have CF. They put the normal gene inside an epithelial cell from a CF patient. The CF cell began to secrete chloride in a normal fashion — the gene had corrected the abnormal chloride channel. Using this technique, scientists successfully “cured” CF in the test tube.

There are many steps to take gene therapy from the test tube to the patient. Gene therapy, which involves placing normal genes into the airway, is currently under research. Medicines to bypass or assist the poorly functioning CFTR in CF patients are also under active investigation. Early trials have been

Special issues around nutrition

These issues can affect your child’s nutrition:

Excessive salt loss
Children with CF lose extra salt in their sweat. Infants require 1/8th teaspoon of salt per day. In older children, on hot days or when heavy exercise is planned, extra salt should be added to the diet. Offer salty snacks such as pretzels, or ask your CF dietitian for help with this issue.

Infant nutrition
Because babies grow so much, there are often more nutrition problems in the first year of life. You might require frequent help from the dietitian at the beginning. Mothers are encouraged to breastfeed, with the use of enzymes. Sometimes formula are added to pumped breast milk to increase calories. Formulas with forms of protein and fat that are more easily digested and absorbed may be used to supplement breast milk. Standard infant formula may also be used with enzymes.

Lung infections and nutrition
When a child has a lung infection and is coughing more often, his appetite usually decreases. Vomiting from coughing attacks is also common. Your child may prefer small, frequent meals during this period. Try to avoid eating before chest therapy because it may cause vomiting. If your child’s appetite decreases, call your doctor. Your child may need antibiotics for a lung infection. Also, the dietitian can suggest some high-calorie supplements, such as Ensure, Pediasure, Resource, Scandishake or over-the-counter and homemade instant breakfasts and high-calorie snacks to provide extra calories for this period.

Adolescents
Adolescents have increased need for calories, protein, calcium, iron and zinc. Teenagers often eat many meals away from home, so it is important to check often that they are getting those calories and taking enzymes when they eat out.
High-calorie supplements
Due to absorption problems in the intestines and the need for extra calories and protein, special products can often give that extra boost. Many children and adolescents need nutritional supplements despite eating high-calorie meals and snacks. Supplements for extra calories may range from homemade milkshakes to commercial drinks. Some children need tube feedings to supplement nutrition to enable them to gain weight and grow. Ask your dietitian. Here is a list of some high-calorie supplements on the market. We will help you get these if your child needs them:

High-calorie food additives
- Polycose – A pure carbohydrate in an easily mixed powered form. It has no taste or sweetness, but helps add calories to food and drinks without increasing volume (Ross Labs)
- Scandical Powder (Scandipharm)
- Additions Powder, without flavor (Nestle)
- Butter or margarine
- Cream or half-and-half

Ready-to-drink high-calorie drinks
- Scandishakes (Scandipharm)
- Boost, Boost Plus (MeadJohnson)
- Ensure (Ross Labs)
- Ensure Plus – A liquid supplement providing balanced nutrition, with a fat content similar to whole milk, available in a variety of flavors (Ross Labs)
- Instant Breakfast – Mixed with whole milk
- Pediasure – High-calorie balanced supplement for ages 1 to 6 years (Ross Labs)
- Bright Beginnings
- Carnation Instant Breakfast – Very high calorie

Ask the CF clinic dietitian for more recipes and ideas for adding calories to foods and drinks.

Remember: Give your child enzymes along with any of these supplements.

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Cystic fibrosis and genetic basics

Genes and carriers
Genes direct the growth and function of the cells in our body. Each person has around 30,000 pairs of genes in each cell. One copy of each pair comes from each parent. Cystic fibrosis is an autosomal recessive condition. This means that both genes of the pair must have a mutation abnormality to have the condition. About 1 in every 28 Caucasian persons has a mutation in one of their two CFTR genes: they are CF “carriers” and healthy. It is possible, but less common, for CF carriers to be of other ethnicities. Carriers are only identified if they have a relative with CF or if specialized testing has been performed to detect a CFTR gene mutation. When both partners are CF carriers there is a 25% chance for each child to inherit two abnormal CFTR genes and have CF.

Genetic prenatal testing
When a child has cystic fibrosis, the child’s brothers and sisters, aunts and uncles, and other family members can be tested to find out if they carry a CFTR gene mutation. The child with CF is first tested to identify the specific gene mutations for that family. Carriers may want to consider prenatal testing for CF in their pregnancies. Preimplantation genetic diagnosis is also an option that uses in vitro fertilization to create pregnancies without the family CFTR mutations. These procedures are accurate; however, they require testing of family members. It is best if carrier testing is performed before a couple is pregnant.

Genetic counseling
A genetic counselor is available at Children’s to discuss questions and concerns about the genetics of cystic fibrosis. To make an appointment to meet with the genetic counselor, call the Genetics Clinic at 206-987-2865. The genetic counselor sees families the same day as Cystic Fibrosis Clinic or on a separate day if that is more convenient.

During genetic counseling, a complete family history is taken and reviewed. The session includes a discussion of the genetics of cystic fibrosis, implications for the family members and a review of available genetic testing.

Genetic counseling and testing services may also be closer to home in your own community. The genetic counselor at Children’s would be happy to provide you with information about these resources.
Cystic Fibrosis Parent Handbook

Care of the lungs

Airway Clearance Techniques (ACT)

Taking care of the lungs is an important part of living with CF. Children with CF have increased mucus (sputum) in their lungs. This mucus is dehydrated, so it is thick and hard to clear. A key part of lung therapy is removing this mucus. Airway Clearance Techniques (ACT) are treatments that loosen thick, sticky lung mucus so it can be cleared by coughing or huffing. Clearing the airways reduces lung infections and improves lung function. We recommend learning the techniques early so both you and your child become comfortable with them. We will introduce ACT to you within the first 6 months after diagnosis.

There are many kinds of ACT like trained coughing, huffing, breathing exercises and clapping on the chest wall, and other forms like the use of breathing devices and special oscillating vests. Most are easy to do. As children with CF grow older, they can do more kinds of ACT. You will work with the respiratory therapists in CF clinic to find the ACT techniques that work best.

Coughing

One of nature's ways of bringing up the mucus is coughing. Coughing is the most basic ACT. Children with CF should not suppress coughing, and should learn proper coughing/huffing techniques to clear mucus from the smaller airways. Huffing is a type of cough. It involves taking a breath in and actively exhaling. It is more like "huffing" onto a mirror or window to steam it up.

Chest Physical Therapy

Chest physical therapy (chest PT or CPT) is often the first ACT taught to families in CF clinic. It is a combination of clapping or vibrating the chest wall (chest percussion) and placing the child in different positions (postural drainage). This helps to drain mucus from different parts of the lungs and aids in bringing the mucus up to be coughed out. The combination of chest percussion with postural drainage is called chest physical therapy.

My daily schedule — Master (to copy)

<table>
<thead>
<tr>
<th>Time</th>
<th>Activity (food, meds, airway clearance, and other daily activities)</th>
</tr>
</thead>
<tbody>
<tr>
<td>6:00 am</td>
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<tr>
<td>7:00 am</td>
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<td>8:00 am</td>
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<td>10:00 am</td>
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<td>12:00 pm</td>
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<td>1:00 pm</td>
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<td>2:00 pm</td>
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<td>3:00 pm</td>
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<td>4:00 pm</td>
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<td>6:00 pm</td>
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<td>10:00 pm</td>
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<td>11:00 pm</td>
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<tr>
<td>12:00 am</td>
<td></td>
</tr>
</tbody>
</table>
If your child is under the age of 3 years, or has gastroesophageal reflux, do not do any chest physiotherapy with their head lower than their body. Gastroesophageal reflux (GE reflux) is when food and acid from your baby’s stomach come back up into the esophagus. These children should use the “modified positions” for CPT. After age 3 years, children who do not have gastroesophageal reflux can use the “standard positions” for CPT. Please see the next pages for both the modified and standard CPT positions.

There is no set rule on how often to clap your child, but a routine should be established so that chest physical therapy is done regularly. We suggest once or twice a day at a time that works best for your family. When your child is feeling sick, you may want to increase to three to four times a day.

The technique of chest physical therapy is difficult to describe in writing and can be learned more easily by seeing it done. It is important that you be instructed by a respiratory therapist. There is a therapist at CF Clinic, or make an appointment by calling 206-987-2258.

We will teach you a lot about proper clapping and positioning techniques. Be sure to get an update on your skills every year. As a child grows, the clapping and draining positions change. By the age of 3 to 4 years your child can begin to learn some basic clearing methods on their own, such as belly breathing and huffing.

### Appendix

#### My daily schedule — Sample

<table>
<thead>
<tr>
<th>Time</th>
<th>Activity (food, meds, airway clearance, and other daily activities)</th>
</tr>
</thead>
<tbody>
<tr>
<td>6:00 am</td>
<td></td>
</tr>
<tr>
<td>7:00 am</td>
<td>Wake up, Nebs, ACT, Breakfast with enzymes, vitamins</td>
</tr>
<tr>
<td>8:00 am</td>
<td></td>
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<tr>
<td>9:00 am</td>
<td></td>
</tr>
<tr>
<td>10:00 am</td>
<td>School snack with enzymes</td>
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<tr>
<td>11:00 am</td>
<td></td>
</tr>
<tr>
<td>12:00 pm</td>
<td>Lunch with enzymes</td>
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<tr>
<td>1:00 pm</td>
<td></td>
</tr>
<tr>
<td>2:00 pm</td>
<td></td>
</tr>
<tr>
<td>3:00 pm</td>
<td>After-school snack with enzymes</td>
</tr>
<tr>
<td>4:00 pm</td>
<td></td>
</tr>
<tr>
<td>5:00 pm</td>
<td></td>
</tr>
<tr>
<td>6:00 pm</td>
<td>Dinner with enzymes</td>
</tr>
<tr>
<td>7:00 pm</td>
<td></td>
</tr>
<tr>
<td>8:00 pm</td>
<td>Nebs, ACT</td>
</tr>
<tr>
<td>9:00 pm</td>
<td>Bedtime snack with enzymes</td>
</tr>
<tr>
<td>10:00 pm</td>
<td>Bedtime</td>
</tr>
</tbody>
</table>
A look at the lungs and percussion techniques

Tracheobronchial tree
a simple look at the airways and lobes of the lungs

Ways to percuss (clap)

For small children, use a CPT cup (percussor)
For older children, use cupped hands.

CPT cup

Phone numbers for key CF Team Members

- Cystic fibrosis nurses 206-987-3316
- Dietitian 206-987-1744
- Respiratory therapists 206-987-2258
- Scheduling (family service coordinator) 206-987-5610
- Social worker 206-987-4144
- Clinic coordinator/program assistant 206-987-2024
- Research nurses 206-987-3921

- **Medical assistants** will bring patients and families to their rooms in clinic and take your child's height, weight and vital signs.
- **CF doctors** provide direct patient care and coordinate care with other health team members. They communicate regularly with your primary doctor to assure continuity of care for the patient with cystic fibrosis.
- **CF nurses** meet with you and your child to discuss concerns and questions, and to provide educational materials. They make referrals, if needed, to outside agencies and provide support and follow-up care. If you have questions after your clinic visit or you believe your child needs medical care, call the CF nurses during regular office hours.
- **CF research nurses** describe current research studies for you and your child -- they may see you in clinic or at separate research visits.
- The **dietitian** checks whether your child is well nourished. They may obtain a three-day food record and measure the patient's arm to determine fat and muscle stores. They suggest special foods to increase calorie intake and instruct patients about the use of enzymes and vitamins. They can also advise about the use of dietary supplements.
- **Genetic counselors** explain the inheritance of CF to parents, family members and older patients. They can update families on the latest research in the areas of prenatal diagnosis and carrier testing.
- **Respiratory therapists** teach families airway clearance techniques and chest physical therapy. If special equipment such as nebulizers, Flutter, Acapella, Phlegm Fighters, or the Vest is prescribed by the doctor, the respiratory therapist will teach proper use of these devices.
- **Social worker** provides information and advice to families about insurance coverage or financial assistance. They provide short-term counseling for both patients and parents to help cope with the stresses of a chronic illness. They are advocates in the community to help families educate teachers and employers about the special needs of people who have CF.
- **Parents** are the most important part of the team. You know your child best. You see your child every day. You will be the person to notice small changes before they become big problems.
- **Your child.** As your child gets older, he or she will become part of the team too, especially during early adolescence. During the pre-teen or teen years your child will be able to take on more of their own care and will be encouraged to call the clinic to schedule appointments and ask questions or voice concerns as they come up.

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Collarbone (clavicle)
Trachea
Upper Lobe
Right Middle Lobe
Lingula (left upper lobe area)
Lower Lobe
Diaphragm

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Cystic Fibrosis Parent Handbook

Chest Physical Therapy (CPT) Infant/Modified Positions

Follow these instructions daily to clear the mucus from your baby’s lungs.

To clear the upper lobes of the lungs’ front upper segments, right and left side
Percuss or vibrate over the shoulder and/or just below the collarbone on both sides of the chest. Keep your child’s spine straight and avoid slumped posture. (See picture at right.) This prevents putting pressure on the stomach, which can cause your baby to spit up.

To clear the lower lobes of the lungs’ back upper and lower segments, right and left side
- Place your child on the stomach with head turned to one side. Child should be lying in a horizontal (flat) position. (See picture at right.)
- Apply percussion or vibration over the back, taking care to avoid the spine.
- Start at the shoulder blades and percuss downward to just above the lower ribs.

To clear the lower lobes of the lungs, front and sides
Place child on right or left side, flat. Start at the armpit and percuss downward to just above the lower rib.

The Cystic Fibrosis Clinic

Coming to your appointments at the Cystic Fibrosis Clinic

How often do we come into the clinic?
Studies suggest that patients with cystic fibrosis who are seen regularly in CF clinic tend to do better than those who don’t have regular visits. For this reason, the CF Foundation recommends quarterly visits (one visit every 3 months). A CF clinic visit usually takes between 1 and 3 hours. While this seems like a long time, it gives your family a chance to meet with many of the different CF team members.

What do we need to know about germ control in the waiting room?
While CF is not contagious, children with CF can share the germs or bacteria that live in their lungs with each other. In CF clinic, we try to reduce the spread of germs by avoiding direct contact between CF patients. When coming in for a clinic visit, please help stop the spread of germs.
- Wash hands frequently.
- Keep your child separate from other children in the waiting room.
- Bring your own toys to clinic rather than playing with the toys in the clinic room.
- Have your child wear a mask while in the waiting areas of clinic. The masks may be removed once you are in your clinic room.

These steps are done to prevent your child from being exposed to germs from others and to protect others from your child’s germs.

Who are the people involved in my child’s care?
The complete care of a child with CF requires a team. You are the most important part of this team. The healthcare professionals you will meet at the CF Center include:
- The clinic coordinator/program assistant organizes the clinic, answers incoming phone calls (Monday through Friday) and keeps current records on all patients.
- The family service coordinator schedules patient appointments and helps schedule referrals.
Chest Physical Therapy (CPT)
Standard Positions

These pictures show a person without a shirt for education purposes only.

Back upper lobes, right and left sides
Place child in a sitting position, leaning forward over a pillow. Percuss or vibrate over the upper back on the right and left sides. Avoid the spine.

Front upper lobes, right and left sides
Have child lie flat on his or her back. Percuss or vibrate between the collarbone and nipple on both right and left sides. Avoid the breastbone.

Back lower lobes, right and left sides
Have child lie on their stomach in head-down position. Percuss or vibrate starting at the shoulder blades and moving down to just above the lower rib. Avoid the spine.

Front and side lower left lobe
Have child lie on their right side in a head-down position. Clap or vibrate over lower rib cage area.

Front and side lower right lobe
Have child lie on their left side in a head-down position. Clap over lower ribs.

Over-the-counter medicines

Never use cough syrups with suppressants on a routine basis. Use of cough suppressants may mask signs of a lung infection needing antibiotic treatment. If your child's coughing is keeping them awake at night, please discuss this with your CF doctor.

Many children with cystic fibrosis have sinusitis, or sinus infections. Confirmed sinus infections are treated with antibiotics and regular nasal steroid sprays to decrease swelling. From time to time, a decongestant is prescribed to reduce the symptoms of stuffiness and constant nasal drainage. Decongestants can cause problems because they may dry secretions. This makes it more difficult to clear the lungs. Your CF doctor should decide whether or not they should be used.

For fevers, over-the-counter drugs containing aspirin should never be used in children, as they may rarely lead to Reye's Syndrome. Medicine containing acetaminophen (Tylenol), or ibuprofen (Advil) should be used instead. In all cases, please consult the CF Clinic if your child's fever lasts for longer than three days.
Other Forms of ACT

Several other forms of ACT are being used across the country and may be of interest to you. These include positive expiratory pressure devices like Flutter and Acapella. When a person with CF blows all the way out through these devices it causes vibration in the airways that helps to dislodge and move mucus. Oscillating vests are another option. These vibrate the chest from the outside to loosen mucus. All of these techniques require training. Our respiratory therapists will be happy to talk with you about which ones (if any) might work for your child and provide you training.

By age 5 to 7 years, your child can begin learning simple independent methods, such as active cycle of breathing, flutter or acapella. By the late teens and early 20s, your child may be ready to leave home, and should be doing ACT by themselves.

If you would like an update in ACT, please ask in CF Clinic or call Respiratory Therapy at 206-987-2258.

Taking antibiotics for lung infections

There are so many antibiotics it is not possible to give you information on every type in this handbook. However, we wanted to let you know about a few commonly used antibiotics. See the table below and on the next page.

Please be aware of any signs of an allergic reaction when starting a new antibiotic. Allergies can develop to antibiotics at any point in time. Signs and symptoms include hives, rash, or swelling of eyes, mouth or tongue. Please call the CF clinic if any of these signs or symptoms becomes present. In the unlikely event that your child has severe symptoms such as shortness of breath or difficulty breathing, call 911.

### Oral antibiotics (taken by mouth)

<table>
<thead>
<tr>
<th>Generic name</th>
<th>Brand names</th>
<th>Side effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amoxicillin and clavulanic acid</td>
<td>Augmentin</td>
<td>• Diarrhea, loose stools</td>
</tr>
<tr>
<td>Azithromycin</td>
<td>Zithromax</td>
<td>• Diarrhea, loose stools</td>
</tr>
<tr>
<td>Cefuroxime</td>
<td>Cefin</td>
<td>• Diarrhea, loose stools</td>
</tr>
<tr>
<td>Cephalexin</td>
<td>Keflex</td>
<td>• Diarrhea, loose stools</td>
</tr>
<tr>
<td>Ciprofloxacin</td>
<td>Cipro</td>
<td>• Sensitivity to sun (wear sunscreen)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Change in mood</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Joint pain or swelling</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Diarrhea, loose stools</td>
</tr>
<tr>
<td>Clarithromycin</td>
<td>Biaxin</td>
<td>• Stomach pain</td>
</tr>
<tr>
<td>Co-trimoxazole (sulfamethoxazole-trimethoprim)</td>
<td>Bactrim, Septra, Bactrim DS, Septra DS</td>
<td>• Anemia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Diarrhea, loose stools</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Rash</td>
</tr>
<tr>
<td></td>
<td></td>
<td>• Sensitivity to sun (wear sunscreen)</td>
</tr>
</tbody>
</table>
Medicines to help clear mucus

Bronchodilators
Some children are prescribed a medicine called a bronchodilator (like albuterol) to use before doing their ACT. These medicines relax the smooth muscles that line the airways. This helps to open the airways. We do not recommend the use of over-the-counter bronchodilators (such as Primatine Mist).

Inhaled steroids
These medicines may be given to prevent symptoms of wheezing and tightness in the lungs. They work by decreasing swelling or inflammation in the airways. Most times, these medicines are inhaled directly into the lungs. A spacer should always be used with metered dose inhalers. A spacer is a tube-like device that fits over the mouthpiece of the inhaler to help you get more medicine into the lungs. In some children a combination medicine is given. This combines a bronchodilator with an inhaled steroid.

DNase
In the mid 1990s, a new medicine called Pulmozyme (DNase) was developed. It is an inhaled preventative medicine that helps to thin the thick mucus produced in the CF lung. A recent study showed it can help slow the progression of lung damage in patients with mild disease.

Hypersal
In 2004, hypertonic saline (Hypersal), was studied and found safe for use in patients with CF. Hypertonic saline works by pulling the natural water from the lungs and rehydrating the sticky, thick mucus, so it is easier to cough out. This medicine may increase lung function and reduce the number of respiratory infections your child has. Pulmonary function tests can help show whether your child would benefit from these types of medicine.

Watching for lung infections (pulmonary exacerbations)

Most lung infections begin with a cold. Symptoms include a runny nose, low-grade fever, and a cough. You should be concerned and call the doctor if your child has any of these signs with their illness:

- a cough lasting longer than 3 to 5 days
- a cough that is linked to vomiting
- weight loss
- loss of appetite
- reduced ability to play or exercise
- fever that lasts
- rattling or wheezing in the chest
- increased or new onset of sputum production

It is hard to predict how often a child will have lung infections — each child is unique. Over time, many kids with CF develop a chronic cough even when they are not sick. Some may cough up some sputum every day. When this happens, it is hard to tell when your child has an acute (short-term) illness like a cold or lung infection. Signs that the cough is related to an illness are:

- changes in activity level
- can’t keep up with friends when playing
- changes in school performance
- lack of appetite, weight loss
- vomiting
- more, thicker sputum that changes from clear to yellow or green in color

If you think your child has a lung infection, increase the frequency of chest physical therapy to three to four times a day and contact the CF Center. We may give an antibiotic.
Exercise is an important part of any child’s healthy growth and development. Children who have cystic fibrosis have a special need for regular exercise and active play. Children, teens and adults with CF who exercise appear to do better than those who don’t. Exercise strengthens the muscles used for breathing. Regular, sustained exercise also improves the way oxygen is used by the lungs, heart and muscles. Aerobic exercise such as bicycling, swimming, or jogging combined with exercise that increases core strength and chest mobility helps the most.

A physical therapist (PT) can work with you and your child to make an exercise plan. They will help pick activities your child enjoys and that allow them to work at their fullest potential. An exercise program promotes greater body awareness and improves self-image through physical achievement. An exercise program probably cannot replace airway clearance techniques; a combination of both is recommended.

Monitoring the lungs

Pulmonary Function Tests (PFTs)

A frequent concern with cystic fibrosis is the amount of lung damage that occurs from repeated lung infections. An exam of the lungs with the stethoscope and chest X-rays gives doctors only a rough estimate of lung damage. At each clinic visit, a respiratory therapist will use a machine called a spirometer to measure your child's lung capacity. This test is called the pulmonary function test (PFT). These breathing tests find out how fast the air can move in and out of the lungs, how much air goes in the lungs after breathing in (inhaling) and how much air is left, or trapped, in the lungs after breathing out (exhaling). The machine can also be used to see if your child responds to bronchodilator medicines or to treatment during an infection.

Pulmonary function tests require coordination and are effort dependent. Your child has to blow very hard into the machine when asked. We have children begin attempts at PFTs at about 4 years of age, but often it is not until age 5 or 6 that children can perform these tests reliably.

Pulmonary function tests are done in the pulmonary function lab or in a room close to clinic. Your child will be coached by a respiratory therapist to take a big breath in and blow all the air out forcefully. They’ll need to repeat the test 2 or 3 times to get an accurate reading.

Oximetry

Oximetry also measures how well the lungs are working at bringing in oxygen to the body’s cells. This test measures the amount of oxygen bound to hemoglobin (a molecule that carries oxygen in red blood cells). It is a painless procedure and involves placing a sensor on either a finger tip or a toe. Oximetry helps doctors find out whether your child needs to receive extra oxygen.