Our Journey™ With Cystic Fibrosis

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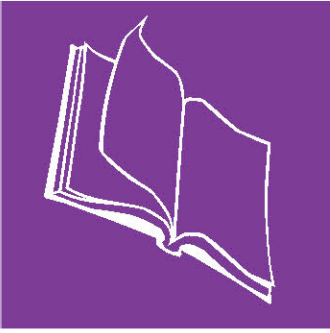
Phoenix Children's Hospital®



The Emily Center



Our Journey with Cystic Fibrosis (CF) Family Handbook

#1869

Phoenix Children's Hospital

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1. I can tell you about cystic fibrosis (CF)

When a child is diagnosed with a serious condition, parents often feel stress and a loss of control because they do not understand everything that is happening. However, once they learn about the illness and master home care skills, parents get their sense of control back.

Perhaps you, too, are feeling stressed and overwhelmed because you need to learn about your child's **cystic fibrosis**. We are here to help. The medical team at Phoenix Children's Hospital follows the hospital's vision to provide hope, healing, and the highest quality care available to your child.

At PCH, we use Journey Boards to help families learn how to safely care for their children. Journey Boards provide support for patient and family education. We will help you become comfortable and confident in your ability to take care of your child with cystic fibrosis by using the Journey Board, **Our Journey with Cystic Fibrosis**.

At Phoenix Children's Hospital, we use Journey Boards to help families learn how to safely care for their children.

What is a Journey Board?

The purpose of a Journey Board is to provide a visual guide for patient and family education. It supports the idea of family-centered care. The Journey Board shows you all the topics we will talk about.

Cystic fibrosis:

Cystic fibrosis (**CF**) is a type of **genetic** disease that can affect different organs in the body. A genetic disease means that it is passed down (**inherited**) from the parents' genes to a child. To be diagnosed with CF, a person must inherit a carrier gene from each of the parents. Many people do not realize the CF gene is in their family until someone finds out they have CF.

CF affects the cells that make mucus, sweat, and digestive secretions. Usually, the body makes thin and slippery secretions. With CF, the body makes secretions that are sticky and thick. These sticky and thick secretions causes mucus to build up in different parts of the body, including the lungs, sinuses, intestines, pancreas, liver, reproductive organs, and the sweat glands.

The **cystic fibrosis transmembrane conductance regulator** (**CFTR**) gene is what provides instructions for making the CFTR protein. The CFTR protein is on the cells that line the lungs, sinuses, intestines, pancreas, liver, reproductive organs, and the sweat glands.

The CFTR protein is responsible for regulating how chloride and water move in and out of these cells.

There are 3 problems that can occur in people with CF:
- no CFTR protein being made
- CFTR protein is made but it does not work correctly or it is not in the right place to work correctly
- CFTR protein is made but not in high enough numbers to work correctly

As a result, chloride and water cannot move in and out of the protein channel, which leads to thick and sticky mucus. This type of mucus can damage many of the body's organs.

How is CF diagnosed?

Diagnosing a person with CF usually takes a few different steps to do. Currently, every person born in the United States goes through **newborn screening**, usually within the first few days of life. This screening helps identify people born with certain types of medical conditions, including CF. Depending on the newborn screening results, a person may need a **sweat chloride test** (also called a **sweat test**) to confirm a diagnosis of CF. The sweat chloride test measures the amount of salt in sweat.

There is no specific cure for CF but your child can still live a full, long, happy life. Your child will need to follow a specific treatment plan to help stay healthy and manage the disease. This includes: following a daily treatment routine, eating well, avoiding germs, and working closely with your child's CF Team.

There is no specific cure for CF but your child can still live a full, long, happy life. Your child will need to follow a specific treatment plan to help stay healthy and manage CF.

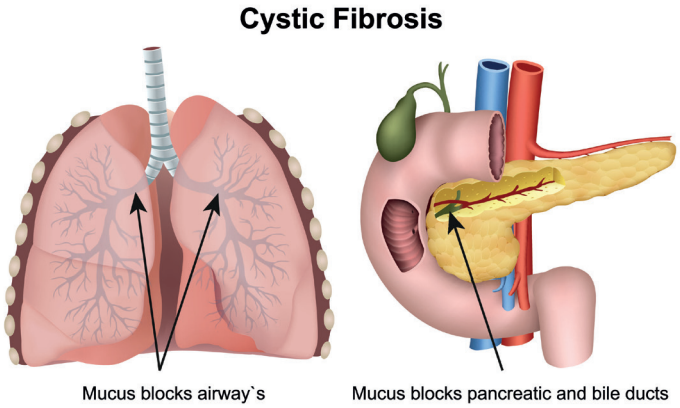
CF can affect the body in different ways:

Sinuses:

- **Sinuses** are air-filled spaces (**cavities**) in the head that are lined with mucus-secreting cells. Sticky, thick secretions can cause sinus blockage, swelling, and frequent infections in the sinuses. Sometimes antibiotics are needed to treat sinus infections or a specialist can provide other interventions such as nasal sinus rinses and nasal sprays.

Lungs:

- The **lungs** are a pair of organs in the chest that remove carbon dioxide and bring oxygen into the blood. Sticky, thick secretions can cause mucus to build up in the lungs. This can lead to swelling and infections and can lead to lung damage.
- Lung damage usually happens slowly over time. Your child will need breathing treatments and airway clearance to help remove mucus from the lungs. Sometimes antibiotics are need to treat lung infections.



In cystic fibrosis, sticky, thick secretions can cause mucus to build up in the body, including in the lungs, pancreas, and bile ducts.

Intestines:

- The **intestines** are part of the digestive system and are a long, continuous tube from the stomach to the bottom (**anus**). The intestines help absorb water and nutrients. Sticky, thick secretions can build up in the intestines. These secretions can cause blockages in the intestines and can cause either loose bowel movements (also called **stools** or poop) or **constipation**. Constipation is when stool moves slowly through the large intestine. It does not come out often enough and comes out hard and dry.
- The secretions can also cause poor absorption of water and nutrients. We will monitor your child's nutrition and stools closely. Your child may need medicine to help keep his or her stools regular.

Pancreas:

- The **pancreas** is a type of gland that is near the stomach. It makes enzymes that go into the small intestine that help with food digestion. The pancreas also makes hormones, including insulin and glucagon, which help control the amount of sugar (**glucose**) in the blood.
- Sticky, thick secretions cause mucus to build up in the pancreatic glands, which can block the release of enzymes needed to digest fats and other nutrients. Your child may need a type of medicine called **pancreatic enzymes** to help the body digest food. These enzymes are usually given before your child eats any meals or snacks. Your child may need to take vitamins as well.
- Some people with CF can develop **cystic fibrosis-related diabetes** (CFRD). This is due to the sticky, thick mucus causing scarring on the pancreas, which prevents the pancreas from producing normal amounts of insulin. It is different from type 1 and type 2 diabetes but has similar characteristics of both types. People with CFRD become **insulin deficient** (similar to someone with type 1 diabetes), which means the pancreas still makes some insulin but not enough to stay healthy. Also with CFRD, people may not respond to insulin in the right way (similar to someone with type 2 diabetes), especially if they are sick, taking certain medicines, or are pregnant. This is called being **insulin resistant**. Both of these things can cause your child to have high blood sugar, or **hyperglycemia**. Common symptoms of hyperglycemia include: increased thirst and urination (peeing). headaches, blurred vision, fruity breath, dry mouth, and trouble concentrating.
- If your child has CFRD, he or she may need to take **insulin** to lower the amount of sugar in your child's blood. Insulin helps maintain healthy blood sugar levels. The amount of insulin your child needs will vary, depending on how many carbohydrates your child eats at meals and snacks.

- Your child may need to see an **endocrinologist** (a doctor that specializes in diabetes). Your child's CF Team together with an endocrinologist will teach you how to calculate how much insulin to give your child.
- Your child will need an **oral glucose tolerance test** (OGTT) to check how well the body's cells can absorb **glucose** (a type of sugar) in a certain amount of time. This test helps diagnose someone with diabetes. This test is done one time a year, starting at age 10 in people with CF who have not been diagnosed with diabetes.

For more information on OGTT, see **Section 4: I can tell you what tests my child is getting and why.**

Liver:

- The **liver** is a solid organ that has many functions in the body. It helps digest food, makes proteins that help the blood clot, helps use and store vitamins, balances hormones in the body, stores food that is used for energy (glycogen), breaks down chemicals and medicines, removes bacteria from the blood and makes immune factors that fight germs, and has many other functions as well.
- In people with CF, thick bile can cause poor bile flow in the liver, which can lead to scarring and **inflammation** (swelling). This is called **cystic fibrosis liver disease** (CFLD). Fatty changes in the liver can also cause scarring, called **steatosis**. This can be due to malnutrition, fatty acid deficiency, or other dietary factors. However, it can also occur in people with excellent nutritional status.
- There are usually no symptoms of CFLD. The liver may be enlarged (**hepatomegaly**) or the spleen may be enlarged (**splenomegaly**) and these are usually found during routine physical exams. Sometimes liver labs are elevated (AST, ALT, alkaline phosphatase, or GGT levels) as well. An ultrasound or liver biopsy can confirm the diagnosis of CFLD. Your child may need to be followed by a **hepatologist** (a doctor who specializes in the liver).

Sweat glands:

- The **sweat glands** are small, tubular glands in the layers of the skin that release sweat onto the surface of the skin. In people who have CF, salt travels with water to the skin and it is not reabsorbed. This can lower the amount of salt in the body, which can cause fatigue, weakness, muscle cramps, dehydration, and other problems.
- Your child may need extra salt to help avoid these types of complications. It also makes the skin salty.

Reproductive system:

- Sticky, thick secretions can cause mucus to build up in the reproductive system for both males and females. This can affect the development and function of the sexual organs, which can make it difficult to have biological children.

For males:

- Most males with CF have an **obstruction** (blockage) or absence of the sperm canal. This is called **congenital bilateral absence of the vas deferens** (CBAVD). The vas deferens is a long tube that carries sperm cells from the **epididymis** (where sperm is stored) to the ejaculatory duct. Sperm production is normal in males with CF but due to the obstruction or blockage, it is harder to fertilize an egg during **intercourse** (sex). Most men with CF can still have biological children but may need to see a **urologist** (a doctor who specializes in the male reproductive system and urinary tract) and use a**ssisted reproductive technology** (ART).

For females:

- Most females with CF have thicker cervical mucus. Females with CF also can have irregular **ovulation** due to poor nutrition. Ovulation is the release of an egg from the **ovary**. The egg travels down the fallopian tube, where fertilization by a sperm cell may happen. Thicker mucus can make it difficult for sperm cells to penetrate the cervix, which may make it harder to become pregnant. Most women with CF can still have biological children but may need to see an **obstetrician** (a doctor who specializes in pregnancy, childbirth, and a female's reproductive system) and use **assisted reproductive technology** (ART).

Now that you have read this:

❒ Tell your nurse or doctor what CF is. (Check when done.)

2. I can tell you what bothers or worries me most about my child's CF

When your child is diagnosed with Cystic Fibrosis, you may be bothered or worried about many things and have a lot of questions. We are here to help you. We want you to feel comfortable in talking with us about your child's diagnosis. We want you to ask questions and we will do everything we can to help you get the right answers. CF is a very complex disease and each child will have different symptoms and follow a different course of the disease. Together, we will help you navigate through your child's individualized treatment plan.

Treatments for CF are constantly changing and improving and it may be hard for you to keep up with the latest information. There may be information you find on the internet that is outdated or incorrect and we encourage you to not look anything up on the internet without guidance from health care professionals. This misinformation may increase your stress. We will give you the correct information and teach you how to care for your child safely.

Your child's CF Team:

Your child's **Cystic Fibrosis Team** (CF Team) includes: a doctor that specializes in CF (**pulmonologist**), a nurse practitioner (NP), a behavioral health doctor (**psychologist**), a nurse, a medical assistant, a respiratory therapist, a nutritionist, a research coordinator, and a social worker. Your child's team is very knowledgeable about the latest treatments and research on CF.

Your child's CF Team is very knowledgeable about the latest treatments and research. They will have recommendations of your child's treatment plan.

The doctors will have recommendations for your child's treatment plans with short-term and long-term goals for your child. The nurses will help you learn how to care for your child safely at home. They are also available to help you sort through any questions you have, such as if you have questions about your child's medicines or if you need to bring your child in for an urgent appointment. Respiratory therapists are experts on your child's breathing treatments and airway clearance treatments. Nutritionists have great ideas for increasing snack and mealtime calories, nutritional supplements, and different meal programs. Social workers have advanced training as counselors and are good listeners. They may connect you to groups and agencies in the community that can help you and your family with finances as well as any mental health concerns. The psychologist on the CF Team is available for preventative help with coping with a chronic disease and can answer questions you may have. He or she is also an ongoing source of support while working through the many demands of CF. All of these team members are available to help you and your child with any questions or concerns you have.

It may also be helpful to have someone else with you during your child's appointments so that you both hear the same information. Then you can talk about it together and may be able to fill in any gaps of knowledge.

Learning about the different medicines and treatments your child will need can feel overwhelming. We know you will have many questions and concerns and we are here to help you. As you learn about your child's CF, we want you to keep asking us questions and any concerns you have until you feel comfortable with your child's care. For example, we want you to be sure you are using the right vest settings for your child since these can change. We also want you to know how to clean your child's equipment correctly since this may change with the latest recommendations.

As your child goes through different phases of CF, you may go through different emotional phases as well. This is normal. For example, when children start kindergarten, their changes in socialization and brain development can lead them to being more aware that not all children do the types of respiratory treatments they do. As they are starting to learn more about CF, they may have more questions about their disease. This could trigger some parents to have feelings of frustration or anger, similar to when their children were first diagnosed with CF. This can be a difficult time for parents and children but we are here to help you on this journey. You and your child are important to us: please talk to us about any feelings or questions you have about CF. We recommend you write down your questions or concerns as you think of them and talk with your health care team about them.

A great resource for information, resources, and articles is the **Cystic Fibrosis Foundation**: https://www.cff.org/

Now that you have read this:

❒ Tell your nurse or doctor what bothers or worries you most about your child's CF. (Check when done.)

3. I can tell you how to comfort my child

When your child has CF, there are many treatments that will become part of your child's daily routine. The best way to approach these treatments is by using the same, calm manner and be consistent. For example, you can approach your child's treatments the same way you approach buckling your child into a car seat. Sometimes, children may not want to be in a car seat, yet you know it is for their health and safety that they always ride in a car seat. By approaching daily treatments for CF in the same manner, it helps children learn what to expect and they will realize treatments are just a regular part of their daily routines.

There are some things that you can do to help your child enjoy and stay comfortable during treatments. To help the vest or nebulizer treatments be more positive, you can do specific activities, play games on the iPad, and watch movies or television (TV) shows only during treatment times to keep these activities special.

There are some things that you can do to help your child enjoy and stay comfortable during treatments. To help the vest or nebulizer treatments, you can do specific activities, such as play games on an iPad.

For example, your child may watch other TV shows during the day, but you can have your child watch a favorite movie during the vest or nebulizer treatment. When you have time, play a special game with your child or read a favorite book during these treatments. The goal is to have your child associate the treatments with positive experiences and make it a special time for your child.

Another way to help your child be more comfortable with his or her treatments is to have your child engage in medical play at home. This can be done with stuffed animals, dolls, brothers and sisters, or with you. For example, your child may pretend to look in someone's throat or do a blood draw with a play syringe or needleless syringe. Medical play can help your child become more calm and relaxed with treatments.

Another way to help your child be more comfortable with his or her treatments is to have your child engage in medical play at home.

Preparing activities for your child to do during CF Clinic visits or hospitalizations can help make these visits a positive experience as well. Bring some of your child's toys, books, coloring books, and crayons when you come to the clinic or hospital. It is a good idea to bring special activities just for clinic visits or hospitalizations, such as a specific coloring book for your child to use. Some children like to have their special blanket or washable stuffed animal with them to make them feel safe and comfortable. Try doing a favorite activity, such as playing an iPad game or reading a pop-up book, during treatments that may be difficult or painful. This may include doing throat swabs or needle sticks.

A way to help your child during a throat swabs is to have your child pant like a puppy, which makes it easier to get the throat swab if you child is doing this. Your child can practice this at home and you can make it fun. Please feel free to bring snacks and drinks to clinic visits and the hospital to help your child feel more comfortable.

Another way to help your child be comfortable is by validating his or her feelings and using coping words. For example, you can teach your child to describe his or her feelings after a procedure, such as saying something is uncomfortable. Then you can tell your child how great he or she did by holding still so it could be done quickly. Praise your child for doing a good job even though the procedure was uncomfortable.

For vests and nebulizer treatments at home, encourage your child to help problem solve different ways to make the treatments more comfortable. Even four year old children appreciate giving their opinions on ways they think would be fun or helpful, even if you already suggested the same thing. Remember, your child does not have the option of whether or not to do his or her treatments or medicines but he or she can suggest ways to make it easier or more fun.

When your child is having difficulty in some areas of treatment, try not to discuss that in front of your child. Instead, focus on the instance in which your child did something well. Have your child overhear you telling others the great things he or she is doing. For example, tell the CF Team when your child did his or her breathing treatment right away that morning. Tell your friend, in front of your child, what a great dinner your child ate last night. If a child has a difficult time doing something, such as a blood draw, research shows that the child will do better for the next blood draw if someone talks to him or her about what went well. Even if it took five people to help your child hold still during a blood draw, talk to your child about how he or she did a great job holding still. With this positive focus, each time will become better and easier.

Take care of yourself:

It is also important to take some time for yourself and to care for yourself. This can help refresh you and give you more energy to take care of your child. Taking care of someone with a long-term disease can be very stressful and can cause someone to have anxiety and depression. Sometimes what happens is the caregiver focuses so much on the child that he or she does not take care of themselves. This can lead to problems that can interfere with daily life.

Signs of anxiety and depression include:

- physical problems, such as stomach or back pain and headaches
- emotional problems, such as anger, sadness, frustration, loneliness, loss of joy, blaming
- mental problems, such as poor attention, confusion, forgetfulness, and mental exhaustion
- spiritual issues, such as feelings of hopelessness

How to help reduce stress:

- When people offer support, let them know how they can help. For example, have someone help you with household chores, such as cleaning and laundry. This can give you more time to finish other tasks, spend time with your family, or help you get to bed at an earlier time.
- Add exercise to your daily routine. This can help with your overall mood and coping as well. Ask a friend or family member to watch your children so you can go for a walk or go to the gym.
- Make sure you get plenty of sleep since this can help your ability to cope with stresses.
- Spend time with your spouse or significant other. Spending time together can help lower your stress level and help you connect with each other.

These activities can help you reduce your stress, help you cope with different situations, and give you more energy. We can also give you suggestions for support and ways to help you cope. Please let us know how you are feeling and we will do our best to help you become more comfortable in caring for your child.

Now that you have read this:

❒ Tell your nurse or doctor how the team can help your child with his or her feelings about his or her diagnosis. (Check when done.)

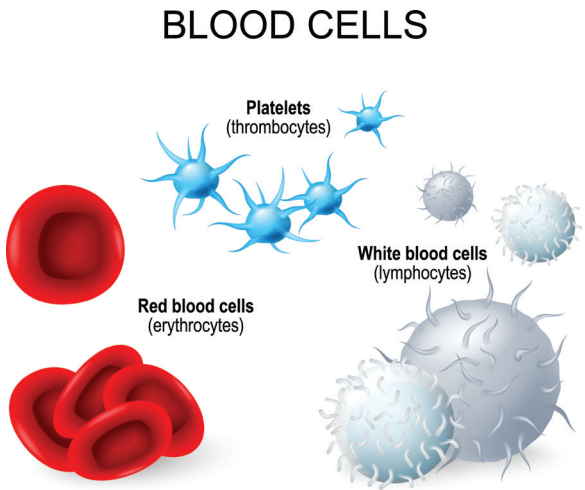
4. I can tell you what tests my child is getting and why

Your child's treatment plan includes doing certain tests on a regular basis, such as collecting blood samples (**blood labs**) and sputum samples. **Sputum** is mucus your child spits up and is sent to a lab for testing.

Your child's treatment plan may include:

Annual blood labs:

Blood is a mixture of fluid (also called **plasma**) and many types of cells, including **red blood cells, white blood cells,** and **platelets**. The liquid plasma is pale yellow , white blood cells and platelets are white, and red blood cells are red. All the cells in the blood are made in the **bone marrow** and start off as **stem cells**.



Blood is a mixture of fluid and many types of cells, including red blood cells, white blood cells, and platelets.

A sample of your child's blood is taken at least one time a year and sent to a lab. The lab runs several types of tests on your child's blood, including:

Complete blood count (CBC):

- Checks blood levels for infection (**high white blood cell count**), anemia (**low red blood cell count**), and other issues.

Comprehensive metabolic panel (CMP):

- Checks electrolyte levels. This is important since people with CF lose more salt and chloride in their sweat than people without CF.
- Checks kidney function.
- Checks for evidence of liver disease caused by CF.

Gamma GT (GGT):

- Checks for liver disease or bile duct problems caused by CF.

Prothrombin (PT), **partial thromboplastin time** (PTT), and **international normalized ratio** (INR):

- Checks how quickly blood clots (also called **blood coagulation**). These labs also measure the Vitamin K level. Vitamin K is needed for normal clotting function. Vitamin K is a fat soluble vitamin and people with CF are at risk of having low vitamin K levels.

Immunoglobulin E (IgE):

- Checks the level of IgE, which is a type of **antibody**. Antibodies are made by the immune system to help protect the body from different types of bacteria, viruses, and allergens.
- If your child's IgE levels are high, your child may have allergies or **allergic bronchopulmonary aspergillosis** (**ABPA**), which is a hypersensitive reaction to the fungus **Aspergillus**.

Vitamin A, D, and E levels:

- Checks the levels of vitamins A, D, and E. These are **fat-soluble vitamins**, which can be low in people with CF since fat is not absorbed (**fat malabsorption**) easily in the body.

Hemoglobin A1C:

- Checks the average level of blood sugar over the past 2-3 months. This is important to monitor for **CF related diabetes** (CFRD).

If your child has any abnormal results from the blood tests, your child may need to repeat the test.

Oral glucose tolerance test (OGTT):

An oral glucose tolerance test checks how well the body's cells can absorb **glucose** (a type of sugar) in a certain amount of time. This test helps diagnose someone with diabetes.

This test is done one time a year, starting at age 10 in people with CF who have not been diagnosed with diabetes. If there are symptoms for diabetes, the test may be ordered at an earlier age.

- Your child will have a **fasting blood sugar** check before the start of the test. This means your child cannot have anything to eat for drink for 8 hours except for water.
- Your child's blood sugar is re-checked 2 hours after drinking a sugary drink.
- The results can determine if your child has CF related diabetes.

Sputum or throat culture:

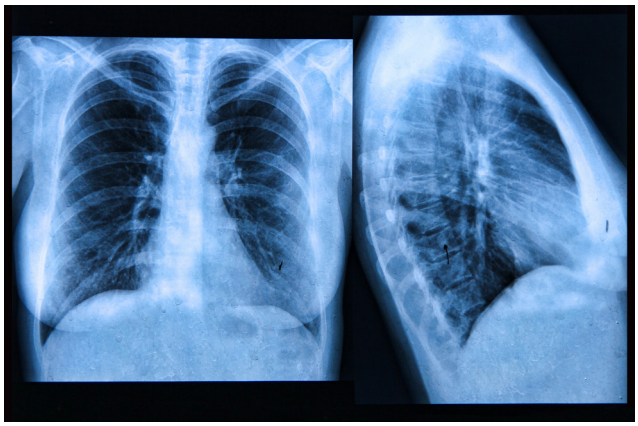
People with CF can have temporary or chronic lung infections caused by different bacteria. Your child needs a sputum or throat culture at every clinic visit. If your child had a culture done recently in the hospital, the emergency room, or at a clinic visit, it will not be re-done.

Your child will need to spit mucus into a cup after coughing or have a **throat swab** done. For the throat swab, a nurse or doctor will quickly move a cotton swab around the back of the mouth to collect a sample. The sample is then sent to the lab to check for germs, such as bacteria, that are growing in the lungs. Depending on the results, your child may need to go on antibiotics to fight the infection.

- There are at least 2 types of bacteria that we will always treat the first time that it is found in a sputum or throat culture: **pseudomonas aeruginosa** (PSA) and **burkholderia cepacia** (B. Cepacia). These are treated even if your child does not show any symptoms of an infection. Most other bacteria are only treated based on symptoms or changes in your child's lung function.
- There are some limitations on sputum or throat cultures. There can be contamination from the nose and mouth, which is normally colonized with bacteria in everyone. **Colonization** means germs are on or in the body but do not make you sick.

Chest x-ray:

- Done at least every 2 years and more often if needed.
- Two pictures are taken for a chest x-ray: one picture from the front of your child's chest and one picture taken from the side.
- Looks for signs of infection or lung damage related to CF.



Chest x-rays are done at least every 2 years and two pictures are taken: one picture from the front of your child's chest and one picture from the side.

Stool elastase:

- Done at the time of diagnosis and may be repeated as needed.
- Tests for pancreatic insufficiency. If the level is low, your child does not have enough enzymes to absorb food appropriately and will need pancreatic enzymes.

Sweat chloride test:

- Done at the time of diagnosis and may need to be repeated if the results are not clear.
- It is the gold standard test done to diagnose cystic fibrosis.
- It tests how the **cystic fibrosis transmembrane regulator** (CFTR) protein functions, which is the primary abnormality in CF.

Pulmonary function tests:

Pulmonary function tests (PFTs) are also called breathing tests. They measure how well your child can move air in and out of the lungs. They can also help find out what is causing any lung problems and can help your child's CF Team develop a treatment plan.

Most children can do PFTs by the age of 6 years old. However, we may start doing PFTs when your child is 4 years old. It can take several clinic visits for your child to get accurate results of a PFT. PFTs are usually done at every visit to the CF clinic.

If your child is short of breath or has had a recent **pneumothorax,** PFTs will not be done during your child's visit to the CF Clinic. A pneumothorax is when air or gas leaks into the space between the lung and chest wall, causing the lung to collapse.

It is best to tell your child what to expect before doing PFTs. Let your child know this test does not hurt and you will stay with your child during it. It may help to make a game out of doing PFTs, such as encouraging your child to pretend to blow out birthday candles or pretend to blow up a balloon when doing PFTs.

Before the test, your child's height and weight are measured. These are used to compare your child's results with the expected lung function of someone with healthy lungs who is similar to your child in age, gender, height, weight, and race or ethnicity.

A specially trained person, usually a respiratory therapist or nurse, will conduct your child's PFTs:

- During the test, your child will wear a soft nose clip so your child will breathe through the mouth only instead of the nose.
- Your child will use a mouthpiece to breathe into (**inhale**) and out of (**exhales**). It is attached to a computer that measures how fast and hard your child exhales.
- Your child will repeat this test a few times to make sure the results are the same.
- Your child may need to take a type of medicine called a **bronchodilator**, which helps relax the muscles in the airway and increases air flow to the lungs. Your child's PFTs are checked again after taking this medicine to see if it helps improve your child's breathing. **Albuterol** and **levalbuterol** are examples of bronchodilators.

For more information, ask your doctor or nurse for the handouts:

- **Chest X-Ray #1753**
- **Sweat Test #2056**

Now that you have read this:

❒ Tell your nurse or doctor what tests your child needs and why they are done. (Check when done.)

5. I can tell you about airway clearance

People with CF have very thick, sticky mucus in their lungs. They usually have problems with lung function and it can cause problems with breathing. It is difficult for them to clear mucus from their lungs just by coughing. **Airway clearance techniques** are useful to help move mucus out of the lungs and they can help maintain the highest lung function possible. If mucus remains in the lungs, it can make it harder to breathe and it can allow germs to grow and cause infections.

Airway clearance techniques are usually started soon after diagnosis of CF because they are be an important part of daily treatments to help keep your child healthy. There are several different types of airway clearance techniques. Most people will use different methods at different times or sometimes a combination of methods to help decrease lung infections and help the lungs work better.

Your child will need to do airway clearance techniques at least twice a day and more often if your child is sick (up to four times a day).

One technique for airway clearance is **chest physiotherapy**, or **CPT**. CPT uses **percussion** (**clapping** or **vibration**) on the chest along with **postural drainage** to help loosen and move mucus out of the lungs. Postural drainage is when a person lies or sits in different positions to help drain mucus from the lungs. A cupped hand or a percussor is used to make a popping sound while clapping on the chest. The sound of the clapping (vibration) helps to break up the mucus. Clapping while in different positions helps drain the loosened mucus from different areas in the lungs.

CPT is usually the first technique taught for airway clearance. A person at any age can do CPT. Most children over one year of age will begin airway clearance techniques using a **high-frequency chest wall oscillation vest**. This is usually called a **vest** or **vest treatment**. The vest looks like an inflatable life-preserver jacket and attaches to a machine that creates high-frequency vibrations that helps loosen and thin mucus. The vest quickly inflates and deflates with air, which creates pressure on the chest, similar to clapping. The vest usually takes place of doing CPT and can provide a more consistent type of airway clearance. It also allows for some independence in doing treatments on his or her own.

Another airway clearance technique is **positive expiratory pressure** (PEP) **therapy**. With PEP therapy, a person uses a device that makes breathing out harder, causing positive pressure in the airways and helps keep the airways open. By forcing the airways open, it can make it easier to get mucus out of the lungs. A **PEP valve** is an example of a type of PEP device that uses resistance when breathing out to help create positive pressure and can make your child cough.

Oscillating PEP devices are other types of devices used in PEP therapy and they work in 2 different ways. These devices use resistance when a person breathes out through them. They also create vibrations when breathing out. These devices work by creating vibrations that are transmitted to the chest when the child breathes out, which is why it is harder to breathe out. The vibrations loosen mucus so it is then easier to cough the mucus out. Both of these ways help move mucus out of the lungs and into the surface of the airways. Then, a person can cough the mucus out. Some examples of oscillating PEP devices are: AerobikA®, Acapella®, and Flutter®.

To get the most benefit from airway clearance techniques, your child needs to do **huff coughs** afterwards. A huff cough is like a mini cough and helps to move mucus that is deep in the lungs. Your child should cough into a tissue or inner elbow to help prevent germs from spreading. Make sure your child cleans his or her hands after coughing by washing hands or using an alcohol-based hand gel.

Your child should cough into a tissue or inner elbow to help prevent germs from spreading.

To do a huff cough:

1. Have your child sit straight up with the chin tilted slightly up.
2. Have your child breathe in a slow but bigger breath than normal and hold it for two to three seconds.
3. Then, keeping the mouth open, have your child forcefully blow out slowly, as if he or she were trying to fog up a mirror across the room. Some people say the word huff when they blow out.
4. After 3 or 4 huff coughs, have your child do a regular hard cough to help clear out the loosened mucus.

For more information, ask your child's nurse or doctor for the handouts:

- **Good Handwashing #100.**
- **How to do Chest Physiotherapy (CPT) #2058**
- **PEP Therapy #202**

Now that you have read this:

❒ Tell your nurse or doctor which types of airway clearance techniques will do, and how to do them. (Check when done.)

6. I can tell you about my child's nutrition plan

There are 3 major nutrients in foods: fats, proteins, and carbohydrates. Every gram of fat has 9 calories while carbohydrates and proteins each have 4 calories per gram. Fats are found in foods such as butter, oil, whole fat dairy, nuts, seeds, fatty fish (such as salmon), and many other foods. Proteins are found in foods such as meats, poultry, fish, eggs, and dairy. Carbohydrates are found in foods such as fruits, vegetables, juices and other flavored drinks, dairy products, grains, beans, and processed foods such as chips and crackers.

Digestion is the breakdown of food into nutrients that the body can absorb easily. These nutrients include fats, proteins, and carbohydrates. The cells in the body use these nutrients to produce energy, promote a healing process, promote body growth, and help a person gain weight.

In general, people with CF need more calories a day than other people their same age. They often have higher energy, or **calorie**, needs for several reasons:

Pancreatic insufficiency:

- One function of the pancreas is to produce **digestive enzymes**, which are then passed through the pancreatic duct to the small intestine to help digest food. In CF, the pancreatic duct can be filled with mucus plugs that can block release of these digestive enzymes and cause food not to be absorbed (**malabsorption**). This causes the body to not be able to digest fat, proteins, and carbohydrates as easily.

Increased work of breathing:

- When a person has an increased work of breathing, the body burns more calories.

Decreased appetite:

- Infections can cause a decreased appetite and a decreased amount of oral intake. For example, when someone has a lung infection, his or her appetite may decrease and nausea may increase, which can lead to a decreased amount of oral intake. Due to this appetite decrease as well as increased energy needs, there can be significant weight loss.

To support exercise:

- A person who exercises, plays sports, dances, or other does other physical activities needs more calories. If a person does not eat enough calories, he or she will lose weight.

Combined, these can make it difficult for a person with CF to gain weight and can also cause unwanted weight loss.

How is my child's growth monitored?

A recommended diet for people with CF is a high calorie, high fat, high protein diet.

Maintaining a healthy weight can also help slow down a person's decline in lung function, which can help a person with CF live a longer and healthier life. The CF Team will take your child's height and weight at each appointment. They will then chart these measurements on a **clinical growth chart** from the Centers for Disease Control and Prevention (CDC). They will also calculate your child's **body mass index** or BMI (a measurement of body fat based on height and weight). The **Cystic Fibrosis Foundation** (CF Foundation) recommends that children under the age of 21 years old should have a BMI at or above the 50% on the growth chart. They also recommend women maintain a BMI of at least 22 and men maintain a BMI of at least 23.

What type of diet should my child eat?

A recommended diet for people with CF is a **high calorie, high fat, high protein diet.** People with CF often have increased caloric needs. If they are also pancreatic insufficient, they have some level of nutrient malabsorption as well.

A well-balanced diet is very important for someone with CF. Eating a well-balanced diet with extra calories and nutrients helps to increase energy for one's physical and emotional well-being. As any condition related to CF worsens, increasing calories is important to help with healing and treatments.

Maintaining a healthy weight can also help slow down a person's decline in lung function, which can help a person with CF live a longer and healthier life. The CF Team will monitor your child's growth closely.

Along with fats, proteins, and carbohydrates, the body also absorbs vitamins and minerals from food. The pancreas plays an important role in the body for the digestion of fat. There are essential vitamins that will only be absorbed from food that contains properly digested fats. These vitamins are called **fat soluble vitamins** and they are vitamins A, D, E, and K.

Along with fats, proteins, and carbohydrates, the body also absorbs vitamins and minerals from food.

Your child may need to take a multi-vitamin that includes all 4 of the fat soluble vitamins. Your child's doctor or dietitian can provide more information if your child needs a multi-vitamin.

Sodium (salt) and chloride levels are monitored closely with CF patients. Your child may need to add salt to meals to help maintain a certain level in the body. Some people with CF must take an oral supplement of salt in order to maintain this level. Your child's doctor or dietitian can provide more information if your child needs more salt.

There are other dietary nutrients that are important to the body as well. These nutrients are called **trace elements** and include, iron, calcium, magnesium, zinc, selenium, and fatty acids. Your child's doctor or dietitian can provide more information if your child has a deficiency in trace elements.

What are digestive enzymes for and when should my child take them?

The pancreas produces digestive enzymes, which help breakdown carbohydrates, proteins, and fats. If a person has pancreatic insufficiency, then he or she may not absorb nutrients from food. This is called **malabsorption**.

Symptoms of malabsorption include:

- Poor weight gain despite a good appetite
- Frequent, loose bowel movements (**stools**)
- Foul smelling bowel movements
- Excessive gas
- Stomach pain
- Bloated stomach (**stomach distention**)
- Oily or greasy stools or oil in the water after stooling

If your child has any of the symptoms of malabsorption, call the CF Clinic.

Pancreatic enzymes (also called **replacement digestive enzymes**) are enzymes that help the body absorb fats, proteins, and complex carbohydrates. **Complex carbohydrates** are made of long-chain sugar molecules. They are found in foods such as whole milk, dairy products (such as cheese), vegetables, peas, beans, and whole grains. **Pancreatic enzyme replacement therapy** (PERT) involves taking the prescribed pancreatic enzymes before eating meals and certain snacks.

Simple carbohydrates are broken down and digested easily in the body. They are found in foods such as fruits, juices, tea, coffee, popsicles, hard candy, gum, and jelly beans. A person does not need to take replacement digestive enzymes when eating only simple carbohydrates.

Pancreatic enzymes usually come in a capsule and do not have a taste. They should be taken just before eating meals and snacks. Older children and adults can swallow the capsules with liquid. The capsule contains beads. For infants and young children, open the capsule and mix the enzymes with applesauce or another acidic food, such as yogurt. It is very important that all the beads are swallowed. It may be hard for infants and young children to take enzymes since they are a different texture than they are used to. As children become more independent, they may refuse to take enzymes. However, it may help to offer them choices on how they would like to take their enzymes. For example, you can ask your child if he or she would like to take the enzymes with applesauce or yogurt.

Pancreatic enzymes are very important to take if someone has CF and is pancreatic insufficient. Most of the time, pancreatic enzymes can treat the symptoms of malabsorption. If a person does not take enzymes before eating, then food cannot be properly digested or absorbed and the body will not receive the necessary nutrients it needs. It can also cause cramping and abdominal pain. By eating a diet high in calories and protein, along with taking the prescribed pancreatic enzymes, it can increase energy and promote weight gain for someone with CF.

Your child's CF doctor will prescribe your child a certain type of pancreatic enzymes. Your child's CF doctor will also tell you how many enzymes to give your child with meals and how many to give with snacks. If there are any questions or problems with your child taking pancreatic enzymes, call your child's CF Team.

Now that you have read this:

- ❒ Tell your nurse or doctor what type of diet your child needs to eat. (Check when done.)
- ❒ Tell your nurse or doctor what pancreatic enzymes are, when your child needs to take them, and how many to take. (Check when done.)

7. I can tell you about my child's medicines and possible side effects

Your child will need to take several types of medicines every day. It is important for you to know what medicines to give your child, why your child needs to take these medicines, when and how to give these medicines, and possible side effects.

Salt:

People with CF have an increased sweat production and causes salt to travel to the skin with water. This salt is not reabsorbed into the body and stays on the skin's surface, which causes the skin to be salty. It also causes lower salt levels in the body. Your child will need salt to replace the lost salt. Although it is plain table salt, it is still an important medicine. Without salt, your child can become **dehydrated** (loss of body fluid that can make the body not work correctly) and it can cause **electrolyte imbalances**. An electrolyte imbalance means that the level of electrolytes in the blood is either too high or too low.

Giving your child too much salt can cause an electrolyte imbalance as well, which can be a side effect. Only give your child the amount of salt ordered by your child's CF Team.

Pancreatic enzymes:

Pancreatic enzymes (also called **replacement digestive enzymes**) are enzymes that help the body absorb fats, proteins, and complex carbohydrates. Complex carbohydrates are made of long-chain sugar molecules. They are found in foods such as whole milk, dairy products (such as cheese), vegetables, peas, beans, and whole grains. **Pancreatic enzyme replacement therapy** (PERT) involves taking the prescribed enzymes before eating meals and snacks.

Simple carbohydrates are broken down and digested easily in the body. They are found in foods such as fruits, juices, tea, coffee, popsicles, hard candy, gum, and jelly beans. A person does not need to take replacement digestive enzymes when eating only simple carbohydrates.

If your child needs pancreatic enzymes and he or she does not take them before eating, it can cause loose stools, stomach pain, cramping, and excessive gas. Your child's body will not absorb the necessary nutrients and calories from eating. This can lead to your child having problems gaining or maintaining weight.

There are no side effects from taking pancreatic enzymes. Store them in a cool, dry place. They should not come in contact with heat.

Bronchodilators:

Bronchodilators are a type of medicine that helps relax the muscles in the airway and increases air flow to the lungs. Albuterol is one type of bronchodilator. Brand names for albuterol include: P**roventil HFA®, Ventolin HFA®, and ProAir®**. Levalbuterol is another type of bronchodilator. The most common brand name is **Xopenex HFA®**. Both types help open up the airways and can help move mucus out of the airways. It can also help with frequent coughing and wheezing. Your child's doctor will tell you how much albuterol to give your child.

Side effects include: fast, pounding or irregular heart rate, shaking (**tremors**), headache, nervousness, and changes in behavior. These side effects usually do not last long after your child takes this medicine. However, if the side effects last more than 1 hour, call your child's CF doctor.

Inhaled corticosteroids:

Some people with CF may have asthma as well. If someone does have asthma, his or her doctor will order **inhaled corticosteroids** (sometimes called ICS). Inhaled corticosteroids help decrease inflammation and decrease mucus production within the lungs. Inhaled corticosteroids are always given with a **mask** if using a machine or with a **spacer** if using an inhaler. Examples include: fluticasone propionate (**Flovent®**) and beclometasone dipropionate (**Qvar®**).

Inhaled corticosteroids are always given with a mask if using a machine or with a spacer if using an inhaler.

Side effects include: dry skin around the mouth, a hoarse voice, and **thrush**. Thrush is a type of yeast infection that can happen in the mouth. It is important for your child to rinse the mouth, wipe the face, and brush teeth after taking this medicine to help side effects. If any of these side effects happen, do not stop taking the medicine and call your child's CF doctor.

Antibiotics:

Antibiotics are a type of medicine to treat bacterial infections. Doctors order antibiotics for people with CF to treat bacteria found in the lungs or throat that may be causing an infection in the lungs. There are different ways to give antibiotics: through a vein (**intravenous** or **IV**), by mouth (**oral**), or inhaled. Your child's doctor orders specific antibiotics based on either a recent culture result or a previous **culture and sensitivities result**. These are types of tests that show which antibiotic works best to fight the infection.

Side effects include: being sick to the stomach (**nausea**), loose stools (**diarrhea**), stomach discomfort, rash, and yeast infections. Some antibiotics can cause ringing in the ears, hearing loss, or problems with balancing. If your child has any of these side effects, call your child's CF doctor right away.

Antibiotics can also kill healthy bacteria in the body and stomach. Your child's CF Team may suggest ways to help prevent and treat side effects from antibiotics.

It is very important that your child finish all of the antibiotics ordered for him or her, even if your child is feeling better. If your child stops taking antibiotics before he or she is supposed to, it can lead to other problems, such as the medicine not killing all the bacteria in your child's body. Then, it can cause the growth of **antibiotic-resistant bacteria**. This means that if the infection comes back, the bacteria may not be killed by taking the same antibiotic.

If your child has to take IV antibiotics for a long period of time, your child may need a **central line**. A central line is an option for long-term access to a vein. The catheter is threaded into one of the large central veins in the chest and goes directly to the heart. There are several types of central lines, including **central venous catheters (CVCs)**, **peripherally inserted central catheters (PICCs)**, and **port-a-caths (ports)**. Your child's CF Team will help decide which type of central line would be best for your child.

Your child will need home health care if your child has a central line at home or is being sent home from the hospital with the port needle left in place. You will be taught how to care for the central line before your child is discharged from the hospital. The CF Team will set up a home care company to provide you with support and supplies for home.

Safety concerns with central lines:

Central lines go directly into the bloodstream. Do not put anything into your child's central line that does not belong in your child's blood. Stop and think before you put anything into the central line. If you put the wrong thing in, like food or medicine meant for the mouth, it could hurt or even kill your child.

Your child's central line has a risk for infection. Make sure you know the **signs of a central line infection**, including:

- drainage, such as oozing or pus
- swelling
- tenderness or soreness
- warmth
- pain
- redness at the exit site
- redness along the catheter path beneath the skin
- a temperature above 101°F (38.3° C) by mouth one time
- a temperature 100.4°F or higher more than one time, taken more than one hour apart by mouth (38° C) (check with your child's health care provider)
- chills
- odor from the exit site

If you notice any of these signs of infection, call your child's CF Team right away or take your child to the Emergency Department.

Dornase alpha:

Dornase alpha is a type of protein (**enzyme**) that helps improve lung function by breaking down thick, sticky mucus in the lungs and makes these secretions thinner. It can also lower the risk for respiratory tract infections. One brand name for dornase alpha is **Pulmozyme®**. It is important to store this medicine in your refrigerator in the foil pouch it comes with. You should not mix this medicine with any other medicine in the nebulizer cup. This could cause the medicines to not work correctly.

Side effects include: chest pain, fever, a hoarse voice, sore throat, rash, and nasal stuffiness. If your child has any of these side effects, call your child's CF doctor.

Hypertonic saline:

Hypertonic saline is a type of inhaled medicine that helps thin mucus in the airways. The mucus is then removed easier when doing airway clearance techniques. It works by increasing the amount of sodium (**salt**) in the airways, which helps thin the mucus and makes it easier to breathe. It is a sterile saline solution and comes in different sodium chloride concentrations. Other names include sodium chloride inhalation and **HyperSal®**.

Side effects include: cough, chest tightness, and sore throat. If your child has any of these side effects, call your child's CF doctor.

Azithromycin:

Azithromycin is a type of antibiotic medicine. However, in people with CF, it is given to help reduce inflammation (**anti-inflammatory**). It is usually given on a regular schedule, such as every Monday, Wednesday, and Friday. Brand names include: **Zithromax®** and **Z-pak®**.

Side effects include: wheezing, being sick to the stomach (**nausea**), and loose stools (**diarrhea**). If your child has any of these side effects, call your child's CF doctor.

Laxatives:

Laxatives are a type of medicine that can help lower the amount of thick mucus and stool in the intestines. In people with CF, mucus and stool can build up in the intestines, causing constipation, abdominal pain, cramping, bloating, and a decrease in appetite. **Constipation** means not having as many bowel movements, or the bowel movements are hard and dry. Laxatives can help by increasing the number of stools and preventing constipation.

There are several different types of laxatives and your child's CF doctor will help you decide which type to give to your child. Examples of laxatives include: polyethylene glycol (**Miralax®**), bisacodyl (**Senna®**), and docusate (**Colace®**).

Side effects depend on what type of laxatives are given but can include: bloating, cramping, being sick to the stomach (**nausea**), and loose stools (**diarrhea**), gas, electrolyte imbalances, and some can interact with other medicines, such as antibiotics and certain heart and bone medicines. If your child has any of these side effects, call your child's CF doctor.

Vitamins:

There are important vitamins that are only absorbed from foods that contain properly digested fats. These vitamins are called **fat soluble vitamins** and they are vitamins A, D, E, and K. They help the body keep working correctly and are stored in fatty tissues and liver. Each type of fat soluble vitamin does different functions in the body.

Vitamin A:

Vitamin A helps the immune system fight infections. It also supports healthy skin, vision, and intestines. Low levels of vitamin A can cause an increased risk for infections, night blindness, hair loss, dry eyes, and skin issues.

Vitamin A is in foods such as eggs, milk, dark colored fruits (peaches, cantaloupes, apricots), and vegetables (broccoli, carrots, spinach, kale).

Vitamin D:

Vitamin D helps build and maintain strong bones and teeth. Low levels of vitamin D can cause thin and brittle bones. People with CF are at risk for developing **osteopenia** and **osteoporosis** (bones are less thick and dense) and have a higher risk for bone breaks. Low levels of vitamin D can also cause weakened muscles, impaired hearing, hair loss, and increased risk for infections.

Vitamin D is produced naturally in the body when the skin is exposed to the sun. However, this may not be enough for the body to work correctly. Vitamin D is in food such as eggs, milk, pudding, fortified cereal, tuna, and salmon.

Vitamin E:

Vitamin E is an **antioxidant**, which means that it helps prevent or slow damage to cells caused by **free radicals**. Low levels of vitamin E can cause muscle weakness, vision issues, trouble walking, and numbness.

Vitamin E is in foods such as almonds, peanuts, hazelnuts, broccoli, whole grain bread, and wheat germ oil.

Vitamin K:

Vitamin K helps the body form blood clots. This is very important so that someone does not bleed to death from a small scrape or scratch. It also helps reduce the risk of heart disease and promotes bone health. Low levels of vitamin K can increase risk for excessive bleeding and also bone fractures.

Vitamin K is in foods such as broccoli, spinach, peas, brussels sprouts, kale, egg yolks, and butter.

Your child may need to take a multi-vitamin that includes all 4 of the fat soluble vitamins. If your child is deficient in other vitamins, he or she may need to take supplements for those vitamins as well. Your child's doctor or dietitian can provide more information if your child needs a multi-vitamin or any other vitamins.

Other medicines:

Your child may need to take other medicines, depending on if your child has any complications from CF. For example, if your child has CFRD, your child may need insulin. Other medicines your child may need include: CF modulators, nasal sprays, gastroesophageal reflux medicines, and **antihistamines** (a type of medicine to help treat allergies). Ask your child's CF Team for more information about these medicines.

For more information, ask your nurse or doctor for the handouts:

- **About Taking Medicines #2067**
- **Always Have Medicine #909**
- **Help Your Child Take Medicine #1116**
- **How to Care for Your Inhaler #103**
- **How to Use Your Inhaler #2060**
- **Measure Medicine in a 1 mL Syringe #721**
- **Measure Medicine in a 3 mL Syringe #2015**
- **Measure Medicine in a 5 mL Syringe #2017**
- **Measure Medicine in a 10 mL Syringe #2019**
- **Medicine List #786**
- **Medicines to Take #337**

Now that you have read this:

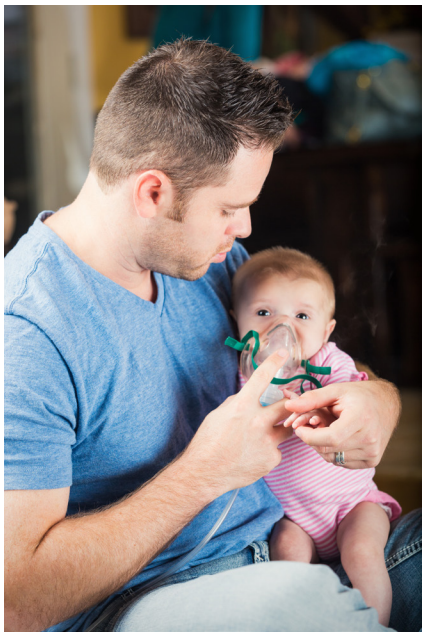
- ❒ Tell your nurse or doctor the names of the medicines you will give your child at home. (Check when done.)
- ❒ Tell your nurse or doctor why each medicine is given. (Check when done.)
- ❒ Tell your nurse or doctor when and how much medicine to give. (Check when done.)
- ❒ Show your nurse or doctor how you would give your child this medicine. (Check when done.)
- ❒ Tell your nurse or doctor if there are medicines or foods your child should not have while taking this medicine. (Check when done.)
- ❒ Tell your nurse or doctor what side effects you will look for. (Check when done.)
- ❒ Tell your nurse or doctor what you would do if you see any of these side effects. (Check when done.)
- ❒ Tell your nurse or doctor where you will fill your child's prescriptions. (Check when done.)
- ❒ Tell your nurse or doctor who you can call if you have a question or concern about a medicine. (Check when done.)

8. I can tell you about my child's home medical equipment and supplies

Nebulizer machines and vest treatments:

CF affects many parts of the body, but a large focus of treatment is on the lungs. Thick and sticky mucus in the lungs can make it hard to breathe. A person's day-to-day living is affected by the severity of the disease. There are many ways to help keep the lungs as healthy as possible. Your child will need special medical equipment and supplies at home.

A **nebulizer** is a type of medical equipment your child will use for inhaled medicines. Nebulizers turn liquid medicine into a mist by using a **nebulizer machine**, also **called a small volume nebulizer** (SVN). Medicine goes into a **nebulizer cup** and then your child breathes in through a mask or mouthpiece. The medicine then travels into your child's lungs.



A nebulizer is a type of medical equipment your child will used for inhaled medicines that turns liquid medicine into a mist.

It is very important to clean and disinfect the nebulizer cup at least once a day. When finished with the nebulizer, it is very important to clean it well. This can help reduce the chance of infection.

The inhaled medicines are usually combined with airway clearance techniques, such as **chest physiotherapy** (CPT) or a high-frequency chest wall oscillation vest. The vest looks like a life-preserver jacket and works by creating high-frequency vibrations that help loosen mucus. The vest takes place of doing CPT and provides a more consistent type of airway clearance. Usually vest treatments take about 20 to 30 minutes. Talk to your child's CF Team if your child's treatments take longer than 30 minutes.

For more information about SVNs and CPT, ask your nurse or doctor for the handouts:

- **How to do Chest Physiotherapy (CPT) #2058**
- **How to Clean a Nebulizer #1055**
- **How to Use a Small Volume Nebulizer (SVN) #2054**

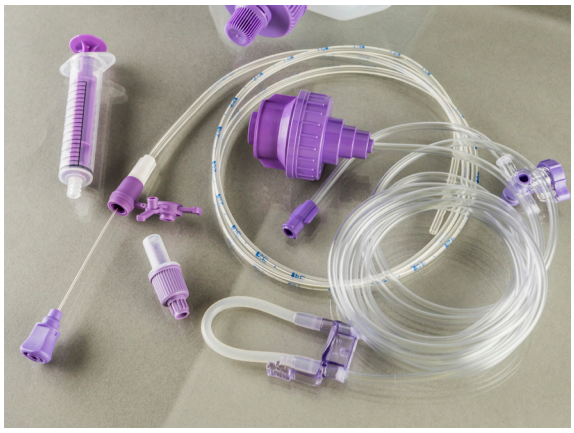
For more information about CPT and vest treatments, see **Section 5: I can tell you about airway clearance.**

Feeding tubes, pumps, and other feeding supplies:

A healthy diet is an important part of helping your child's body grow and work well. The body needs a healthy diet of protein, carbohydrates, fats, vitamins, and minerals. In general, people with CF need more calories a day than other people their same age. If a person cannot eat enough calories each day, he or she may need an **enteral feeding**. An enteral feeding is when food is put through a tube directly into the stomach or small intestine. This food is usually a liquid form of protein, carbohydrates, and fat. It has all the nutrients, vitamins, and minerals to help a child grow and be as healthy as possible.

There are several ways to give an enteral feeding. A **nasogastric tube** is used to put food directly into the stomach. The nasogastric tube is also called an **NG tube**. This tube is put in a nostril and is passed down the throat through the esophagus and into the stomach.

If your child has a **gastrostomy tube (G-tube or GT**), the liquid food or medicine goes directly into your child's stomach. If your child has a **gastro-jejunostomy tube (GJ tube)**, the liquid food or medicine goes directly into the small intestine. Sometimes, a child may get fed through the GJ tube and get medicines through the G-tube.



If your child needs a feeding tube, you will have a home health company that will bring you the supplies you will need for these feeds.

If your child needs a feeding tube, you will have a home health company that will bring you the supplies you need for these feeds.

Supplies for home may include:

- feeding pump
- feeding bag
- extension tubing
- syringes
- formula
- pole to hold feeding bag and feeding pump, if using one

For more information about feeding tubes, ask your nurse or doctor for the handouts:

- **Our Journey with Tube Feeding Journey Board #1882**
- **How to Care for Your Child with a G-Tube (Gastrostomy Tube) or GJ-Tube (Gastro-Jejunostomy Tube) #207**
- **How to Care for Your Child with a Nasogastric (NG) Tube #1824**

Central line supplies:

If your child has to take IV antibiotics for a long period of time, your child may need a **central line**. A central line is an option for long-term access to a vein. The catheter is threaded into one of the large central veins in the chest and goes directly to the heart. There are several types of central lines, including **central venous catheters (CVCs), peripherally inserted central catheters (PICCs), and port-a-caths (ports)**. Your child's CF Team will help decide which type of central line would be best for your child.

Your child will need home health care if your child has a central line at home or is being sent home from the hospital with the port needle left in place. You will be taught how to care for the central line before your child is discharged from the hospital. A home care company will be set up to provide you with support and supplies.

Supplies for home may include:

- saline syringes
- medicine syringes
- heparin syringes (if needed)
- central line dressing change kits
- disinfectant caps
- chlorhexidine with alcohol wipes
- face masks
- clean gloves

For more information about central lines, ask your nurse or doctor for the handouts:

- **How to Care for Your Child's CVC at Home #197**
- **How to Care for Your Child's PICC at Home #426**
- **How to Care for Your Child's Port-a-Cath at Home #810**
- **Our Journey with a Central Line Journey Board #1914**
- **Our Journey with a Central Line App Download Instructions #1963**

Who to call for help:

Your child's home health care company: _____

Your child's home health care nurse: _____

Telephone number: _____

It is a good idea to save this information in your cell phone as well. When you have about two weeks of supplies left, it is time to order more. It may take this much time for the supplies to be approved by your insurance company. If you have any questions or concerns with home supplies, contact the CF Clinic.

Now that you have read this:

- ❒ Tell your nurse or doctor what supplies you need for your child at home. (Check when done.)
- ❒ Tell your nurse or doctor where you will get those supplies and how. (Check when done.)
- ❒ Tell your nurse or doctor who you will call if you have questions or concerns about the supplies. (Check when done.)

9. I can tell you what my child is able to do as he or she continues to develop

Growth and development:

Milestones are also called **stages of development**. Every child's growth and development is different.

Milestones can tell you what to expect from your child in different areas as your child grows, including:

- Physical growth
- Thinking and reasoning (**cognitive development**)
- Emotional and social development
- Language development (**communication**)
- Sensory and motor development

Children usually move in a natural and predictable way from one milestone to the next. However, each child grows and gains skills at his or her own pace. Some children may be advanced in one area, such as language, but behind in another, such as sensory and motor development. If your child is meeting milestones, then his or her development is on track. If your child continually misses milestones or reaches a milestone but then loses that new ability, he or she may need extra help from a doctor or a specialist. Your child's doctor will check for certain milestones at routine checkups. It is also important for you to learn what milestones to watch for.

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Children with CF generally reach developmental milestones at the same time as children who do not have CF. However, physical growth is an area that is frequently affected in children with CF. As a result of the disease, it can be harder to gain weight. This has improved over time with earlier diagnosis of CF and other interventions, such as starting **enteral feeds** (tube feeds) and giving medicines that can help with growth.

Emotional and social development is another area that is sometimes affected by CF. This is because children with CF cannot be in close contact with other children with CF. This is to help prevent spreading certain infections that are more common in children with CF.

The remaining areas of development are generally not affected. Children with CF are able to attend school, participate in sports, and engage in other activities. However, it is important that they keep up with the recommended therapy to help keep their lungs healthy.

For more information, ask your child's doctor or nurse for the handout:

- **What to Expect as Your Healthy Baby Grows: Developmental Milestones #1689**

Physical activity:

All children must play. Even the sickest child can play in some way. Playing helps children understand their world. Many children can relax and forget their worries when they play. Physical activity is greatly beneficial for all children, but it is especially important for children with CF because it can help release mucus from the lungs, strengthen lungs, improve emotional well-being, and slow the rate of your child's disease process. It can also improve your child's immune system, help prevent stress and depression, and can increase the quality of sleep.

Remember, physical activity can be a family or social event. Structured games and playing on a team can encourage teamwork and competitiveness, along with all the other benefits of being physically active. Physical activity may cause more pronounced coughing, which is normal for those with CF. The CF Clinic does not promote high-intensity activities at extreme altitudes, such as mountain climbing. Ask your child's CF doctor what activities your child can do.

Structured games and playing on a team can encourage teamwork and competitiveness, along with all the other benefits of being physically active.

Clubs and other after school activities:

Your child may be interested in participating in clubs and other after school activities. These types of programs can be very beneficial to your child. They can offer ways for your child to interact with others and help him or her become more social. They can help your child grow academically and emotionally. Examples of clubs and after school activities include: performing arts, dance, drama, creative arts, outdoor education (such as Girl or Boy Scouts), and different sports programs. These programs can be affiliated with your child's school, community center, church, library, or nearby park.

Now that you have read this:

❒ Tell your doctor or nurse what your child is able to do. (Check when done.)

10. I can tell you signs my child has a CF exacerbation and how I will respond

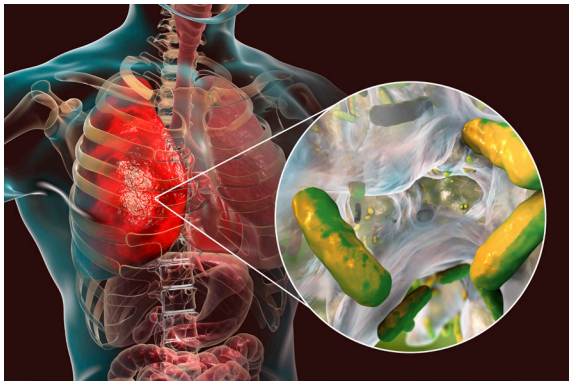
A sudden worsening of symptoms of CF is called an **exacerbation**. This is when symptoms of CF get worse, or flare up, quickly.

You may notice these symptoms with a CF exacerbation:

Respiratory:

With a **respiratory exacerbation**, your child may have:

- chest tightness
- shortness of breath
- coughing
- increased amount of secretions with coughing
- coughing up blood (hemoptysis)
- chest pain
- wheezing
- breathing fast
- labored or difficulty breathing
- sinusitis or nasal polyps
- decreased **pulmonary function test** (PFT)
- a lung infection



With an exacerbation, your child may have a lung infection. Your child may need medicine, such as antibiotics, to kill the bacteria.

Your child may need oxygen to help keep oxygen levels up. Your child may need medicine, such as antibiotics, to kill the bacteria or a steroid to decrease inflammation in the lungs.

Airway clearance is something that needs to be done every day to help your child maintain good lung function. This can help prevent exacerbations. However, if your child is showing symptoms of a respiratory exacerbation, you need to increase the frequency of your child's airway clearance, up to 4 times a day while sick.

Other symptoms of an exacerbation include:

Gastrointestinal:

During an exacerbation, your child may have diarrhea, a decrease or loss of appetite, weight loss, fat in stool, heartburn, severe constipation, or bulky stools. Your child may need a change in enzymes or may need to take medicine to help with heartburn or constipation.

Whole body:

During an exacerbation, your child may get tired more easily (**fatigue**), not be able to exercise like normal, have more salty sweat than normal, and lose weight.

Fever:

Fever can be a symptom of an exacerbation. If your child has an oral fever over 100.4° F (38° C) and it does not go down after taking medicine to treat it, call your child's CF Clinic. If it is outside normal business hours, you may need to take your child to an Urgent Care or the Emergency Department.

Severe exacerbation:

If your child's exacerbation is severe, your child may need to be admitted to Phoenix Children's Hospital. It is common for your child to have a throat swab or sputum culture to find out the type of infection, if there is one. Your child will need more airway clearance therapies and other types of medicines to help with the exacerbation.

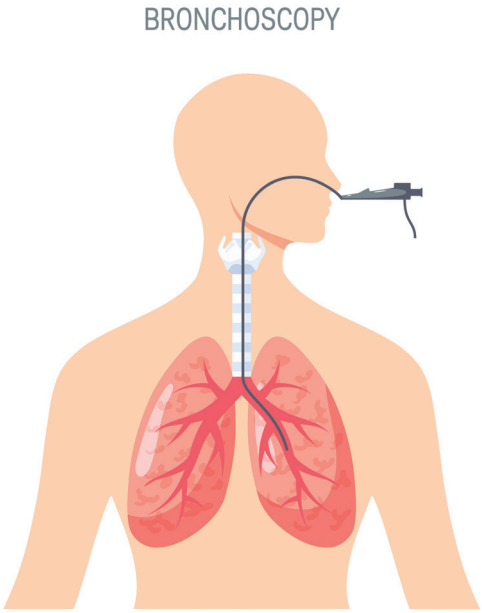
Your child may need an IV for antibiotics. If your child has to take IV antibiotics for a long period of time, your child may need a **central line**. A central line is an option for long-term access to a vein.

For more information about central lines, see **Section 7: I can tell you about my child's medicines and possible side effects.**

Your child may need other tests to see what is causing the exacerbation and these can help determine what treatments your child needs. A sample of your child's blood may be taken to look for infections. Your child may need a **chest x-ray** as well. A chest x-ray is a picture of the lungs, heart, blood vessels, airways, and the bones of the chest and spine. A chest x-ray can show if there is fluid in or around the lungs. It can also show if there is air around the outside of the lungs (called a collapsed lung or **pneumothorax**). Sometimes two pictures are taken for a chest x-ray. These include one picture from the front of your child's chest and one picture taken from the side.

Sometimes, a child may need a **bronchoscopy**. A bronchoscopy looks at the inside of the voice box (**larynx**) and the airways inside the lungs (**bronchi**). This is done with a **flexible fiber optic bronchoscope**. This is a long, thin, bendable tube with a tiny camera and bright light at the end that is used to look inside the lungs. Samples of the liquids in the lungs are taken and sent to the laboratory to look for infections.

Treatment for exacerbations usually takes 2 weeks. Your child may need to stay in the hospital during that time. Sometimes we are able to transition your child home earlier, depending on the care and treatment your child needs.



A bronchoscopy looks at the inside of the voice box and the airways inside the lungs with a flexible fiber optic bronchoscope. Samples of the liquids in the lungs are taken and sent to the laboratory to look for infections.

If your child is admitted to Phoenix Children's Hospital, here are some things you can expect:

- **Team rounds:** Every day, the health care team talks with your CF Team. They will update you about your child's health, test results, and your child's plan of care.
- **Vital signs:** They are a group of tests done to check your child's health status. They are measures of: temperature, blood pressure, heart rate (**pulse**), breathing rate (**respirations**), oxygen saturation (**sats**), and pain. Vital signs are taken at least every 4 hours and more often if needed.

- **Isolation:** Every patient with CF is on **contact and droplet isolation**. This means everyone who enters your child's room must wear a gown, gloves, and mask. This helps keep your child and other patients from picking up germs. Parents and other members of the family may choose not to wear gown, gloves, and masks. However, if they choose not to, they must wash their hands before leaving the room to go to the cafeteria or another common area of the hospital. We also encourage them to use masks when going to common areas, Isolation means the patient must stay in the room at all times. Sometimes it is possible for your child to leave the room and go for a walk. This can only happen with staff members (such as a nurse, child life specialist, or physical therapist) and your child's CF team must say it is okay for your child to do it. During the walk, your child must wear a gown, gloves, and a mask. It can be hard to be in the same room for so long. Ask a child life specialist for activities for your child to do while having to stay in the hospital room.
- **Intake and output:** We keep track of everything your child eats and drinks, and how much urine and stool comes out. It is also called **I & Os**. Do not flush or throw away any pee, poop, vomit, saliva, or any other body fluid.
- **Food and drinks:** You can order meals and snacks for your child through room service and are free of charge for your child.
- **Visitors:** While in the hospital, you and your child may have visitors. Information about visiting hours and overnight guests is in the Phoenix Children's Hospital Admission Booklet. No one under the age of 18 years old can stay after visitor hours are over.
- **1 Darn Cool School:** We have a school at Phoenix Children's Hospital that can help support your child with school work. School is an essential part of your child's normal routine and it is important for your child to keep this routine while in the hospital. Your child's school can also send in school work for your child to help not fall behind while at the hospital. Ask your child's CF Team for more information about 1 Darn Cool School.

For more information, ask your child's doctor or nurse for the handouts:

- **Chest X-Ray #1753**
- **Fever #1691**
- **Flexible Bronchoscopy #2051**
- **Isolation #875**

Now that you have read this:

- ❒ Tell your nurse or doctor the symptoms of a CF exacerbation. (Check when done.)
- ❒ Tell your nurse or doctor what treatment is needed for a severe exacerbation and how long your child may need treatment for it (Check when done.)

11. I can tell you about my child's follow up appointments

People with CF require constant medical care. The Cystic Fibrosis Foundation recommends having appointments with the CF Clinic at least every 3 months and more often when they are sick. The Cystic Fibrosis Foundation recommends children under the age of 2 years be seen more frequently, usually every 1 to 2 weeks at first. Then these visits can be spread out to every 1 to 2 months, depending on how your child is doing. This is so your child's CF Team can closely monitor your child's growth and nutrition levels and make changes to your child's medicine and diet, if needed.

If your child has complications from CF, your child may need more appointments with specialists to help manage these complications. Types of complications that may require more appointments include: feeding tubes (such as an NG tube or G-tube), if your child has a **low body mass index** or BMI (a measurement of body fat based on height and weight), pancreatic insufficiency, frequent sinus or nasal symptoms, or cystic fibrosis related diabetes (CFRD).

Please make sure to keep all of your child's appointments as they are extremely important to your child's health. Please make a follow up appointment before you leave the clinic. If your child cannot make it, please call the CF Clinic and reschedule as soon as possible.

Please make sure to keep all of your child's appointments as they are extremely important to your child's health.

Your child's CF Team includes: a doctor that specializes in CF (**pulmonologist**), a nurse practitioner, a behavioral health doctor (**psychologist)**, a nurse, a medical assistant, a respiratory therapist, a nutritionist, a research coordinator, and a social worker.

Team members will meet with you during your child's appointment to give you information on your child's disease progression and answer any questions. At this appointment, they will give you prescriptions, tests, and referrals, as needed. You will notice the medical team wearing gowns, gloves, and masks when they examine your child. This is to help prevent germs from spreading to different children. You may also see children wearing masks while walking around the hospital to help them keep from getting germs.

Once a year, your child will have an extensive appointment with the CF Team. Plan on being at the CF Clinic for at least 2 hours for this appointment. Your child will have blood work done and a chest x-ray. Have your child wear layered clothing or bring a jacket since it can get cold in the clinic. Feel free to bring food and snacks for your child and bring activities that will keep your child entertained.

If your child needs blood work or other tests, someone from the CF Clinic will let you know so you can plan accordingly.

If your child has a port-a-cath, the CF Team will arrange for a home health nurse to draw blood at home a week or two before your appointment. Make sure to put EMLA® cream on 60 minutes or LMX cream on 20 minute before accessing your child's port-a-cath.

If you have general questions for your child's CF Team, we encourage you to write them down and bring them with you to your child's next clinic appointment.

For more information, ask your nurse or doctor for the handout:

- **You're on the Health Care Team #167**

For more information on a **Clinical Care Schedule** from the Cystic Fibrosis Foundation, see:
https://www.cff.org/For-Caregivers/Clinical-Care-Guidelines/Age-Specific-Clinical-Care-Guidelines/Clinical-Care-Schedule-for-Newborns-to-5-Year-Olds-with-CF.pdf

Now that you have read this:

- ❒ Tell your nurse or doctor how often your child needs to have follow-up appointments. (Check when done.)
- ❒ Tell your nurse or doctor what will happen and who you will see during your child's annual appointment. (Check when done.)

12. I can tell you who I can call with questions or concerns and when I should call

We are here to help you. If you have any questions or concerns on how to care for your child safely at home, call your child's CF Clinic:

If your child is followed at Phoenix Children's Hospital CF Center:

- Monday through Friday from 8:30 a.m. to 4:00 p.m., call the CF Clinic at 602-933-0985 and press Option 3. If the nurse cannot come to the phone, please leave a message. The nurse or another member of the CF Team will call you back as soon as possible.

- If the CF Clinic is closed, call the hospital operator at 602-933-1000 and ask for the PCH on-call pulmonologist.

You may also use the **FollowMyHealth™** portal to access your child's health information at PCH. FollowMyHealth™ is our PCH Patient Portal. It allows you to manage your child's personal medical records 24 hours a day with secure online access from any computer, smartphone, or tablet. Children 13 years old and older can also access their own health records and use all of the tools available in FollowMyHealth™.

If you would like to get started with the FollowMyHealth™ Patient Portal, contact Health Information Management at 602-933-1490 or email: patientportalsupport@phoenixchildrens.com.

Now that you have read this:

- ❒ Tell your nurse or doctor the name and telephone number of your child's doctor. (Check when done.)
- ❒ Tell your nurse or doctor who you should call if you have questions about how to care for your child safely at home. (Check when done.)

If you have any questions or concerns,
❒ call your child's doctor or ❒ call _____

If you want to know more about child health and illness,
visit our library at The Emily Center at Phoenix Children's Hospital
1919 East Thomas Road
Phoenix, AZ 85016
602-933-1400
866-933-6459
www.phoenixchildrens.org
www.theemilycenter.org
Facebook: facebook.com/theemilycenter
Twitter: @emilycenter

January 23, 2020 • In family review
#1869 • Written by PCH CF Team and Nicki Mitchell, MSN, RN, CPN

The Emily Center at Phoenix Children's Hospital

Phoenix Children's Hospital

Si usted desea esta información en español, por favor pídasela a su enfermero o doctor.

#1869

Our Journey with Cystic Fibrosis Family Handbook

Family Review of this Handout

Families: Please let us know what you think of this handout.

Would you say this handout is easy to read? ❒ Yes ❒ No
Please circle the parts of the handout that were hard to understand.

Would you say this handout is interesting to read? ❒ Yes ❒ No
— Why or why not?

Would you do anything differently after reading this handout? ❒ Yes ❒ No
— If yes, what?

After reading this handout, do you have any questions about the subject? ❒ Yes ❒ No
— If yes, what?

Is there anything you do not like about the drawings or pictures? ❒ Yes ❒ No
— If yes, what?

What changes would you make in this handout to make it better or easier to understand?

Please return your review of this handout to your nurse or doctor or send it to the address below. You can also scan it and email it to emilycenter@phoenixchildrens.com.

The Emily Center
Health Education Specialist
Phoenix Children's Hospital
1919 East Thomas Road
Phoenix, AZ 85016-7710