NATIONWIDE CHILDREN'S
When your child needs a hospital, everything matters.℠

Helping Hand™

CYSTIC FIBROSIS

Cystic fibrosis (CF) is an inherited disease. It is not contagious ("catching"). CF affects the digestive system and the glands in the lungs that produce mucus (Picture 1). It also affects the glands that produce sweat and saliva.

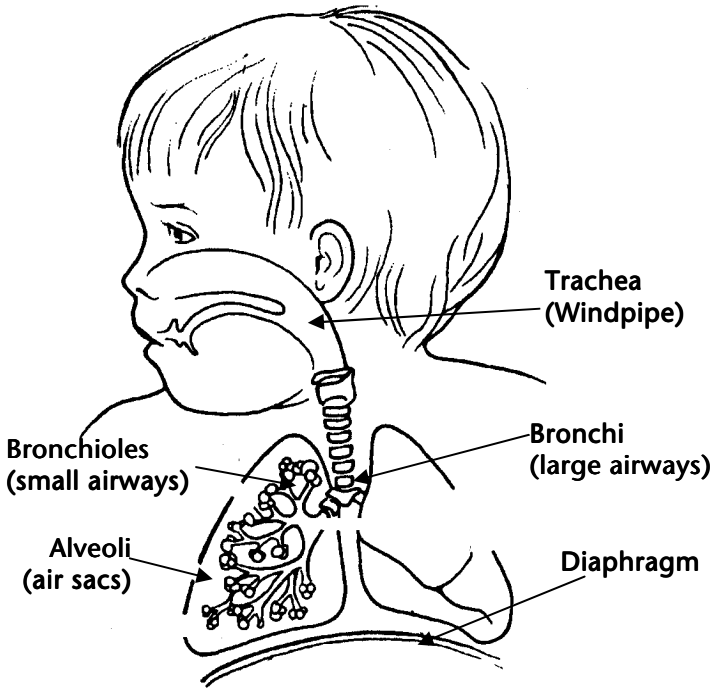
WHAT CAUSES CYSTIC FIBROSIS

Chromosomes (CROW-ma-somes) in our bodies are in every one of our cells. They are what make us different from everyone else. The chromosomes carry the genes that control what we inherit from our family. In cystic fibrosis, the child inherits the gene that causes CF from both parents. In most cases, the parents do not know they carry this gene until their baby is born with CF. Pamphlets that explain the genetic causes of CF are available from the Cystic Fibrosis Foundation. A genetics counselor can also be very helpful to families with a child with cystic fibrosis.

EARLY SIGNS OF CF

- A salty taste on the infant's skin, noticed when kissing the child
- Failure to grow or gain weight (failure to thrive), or weight loss
- Cough that does not go away or comes back often
- Wheezing
- Large, loose, frequent bowel movements (BM), oily BMs, or frothy and foul-smelling BMs
- Chronic sinus infections

A doctor should see a child who has any of these symptoms. Tests can be done to find out if CF is the cause.



Picture 1 The respiratory system inside the body.

TESTING FOR CF

- One of the tests for CF is called a sweat chloride test. A small sample of sweat is collected and the amount of salt in the sweat is measured. It takes about 1 hour to collect a sample. The test is painless. Refer to the Helping Hand: *Sweat Chloride Test,* HH-III-62.
- Tests may also be done to see if there is normal digestion of food in the small intestine. Specific BM tests may be ordered to see if there are problems with food digestion.
- Chest x-rays, lung function tests, and respiratory cultures are other tests that may be done.
- Specific DNA testing for CF is also done by testing a sample of blood.

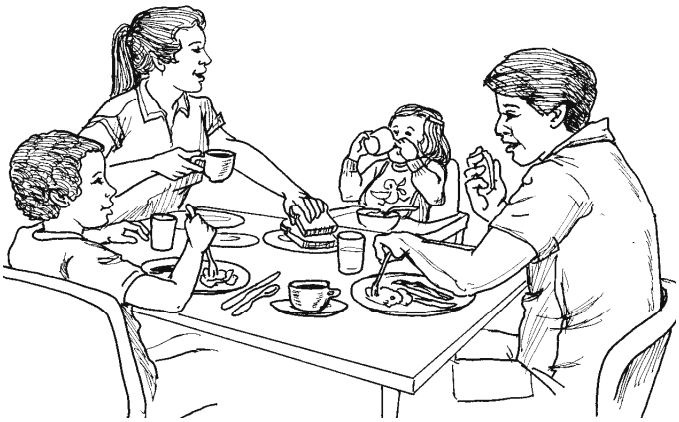
CARE OF THE LUNGS

- One of the most important things you can do to care for your child is to keep the airways of his lungs free of thick mucus. If mucus collects in the lungs, germs can grow in it and cause lung infections.
- Your child may need aerosol breathing treatments to keep his airways moist and the mucus thin. A nurse will instruct you and your child on aerosol treatments.
- Antibiotics are often prescribed for even mild "cold like" symptoms.
- **No one should smoke around your child.**
- Sometimes your child may need to be hospitalized and receive intravenous (IV) antibiotic therapy to help keep his lungs clear.
- Chest physiotherapy (CPT), also called postural drainage, is a treatment used to help move the mucus out of the airways. A nurse will teach you how to give this treatment.
- Other methods of giving CPT are used. Your child's nurse, doctor, or respiratory therapist will help decide which treatment would be the most helpful for your child.
- It is **very important** to complete **all** of the treatments your child's doctor tells you to do every day!

NUTRITION

Your child's doctor and dietitian will help you plan his food needs. The following are general guidelines for children with CF:

- A diet high in calories, protein, vitamins, and minerals is needed for adequate growth and development. Nutrition is especially important for keeping the immune system strong to fight infection. More calories are needed for energy because the lungs work harder to breathe. Extra protein, vitamins, and minerals are needed because they are often lost in the BM.
- We want your child to eat a lot of calories each day. Make sure to talk with his dietitian about ways to help him get enough calories. (Picture 2)
- Enzymes (EN-zimes) are found in the digestive juices. These substances help the body process food. When a child has CF, enzymes made by the pancreas are sometimes blocked and can't reach the small intestine. If this happens, your child's doctor will prescribe enzymes to be taken with meals to help break down and digest food. When enzymes are added to the diet, they help to maintain good nutrition and growth. Enzymes also help the child to have a normal BM.
- Your child will need salt added to the diet to replace the salt that is lost in his sweat. Infants can have salt added directly to the bottle and older children can have it added directly to their food. A dietitian or nurse will review with you the amount of salt needed and teach you how to add it to your child's diet.



Picture 2 Your child should have 3 balanced, high calorie meals and 2 to 3 snacks each day.

- Your child may need vitamin supplements. Your child's doctor will order the diet enzymes and vitamins if your child needs them.
- A dietitian from the Pulmonary Team will spend time talking with you about the foods that will be best for your child.
- You can get recipes from the cystic fibrosis web site (www.cff.org) for high calorie, high fat foods.

PREVENTING INFECTION

- If possible, your child should not be in close contact with people who have colds or other infections, especially as an infant.
- Keep a record of all immunizations and make sure your child gets all booster shots.
- Your child should have a flu shot every year. Two are often required the first year it is given to assure immunity.

ACTIVITY

- Normal play is necessary for your child's development.
- Parents should try not to be over-protective.

SCHOOL

- School-age children should attend school as much as possible. The teacher should be told that your child has CF (ask your nurse for another copy of this Helping Hand). Your child's teacher and other students should be told that CF is **not** contagious. If your child needs to take medicines during school hours, the school nurse and teacher should be told how to give them.
- School nurses and teachers may discuss your child's needs with the nurses on the Pulmonary Team by calling (614) 722-4766. It is important to keep in touch with your child's school.

APPOINTMENTS

- It is important to keep all appointments. If for any reason you cannot keep an appointment, call the Cystic Fibrosis Center at (614) 722-4766 and reschedule.
- Your child will be seen in the CF clinic about every 4 to 8 weeks, and as needed.
- Between appointments, doctors and nurses from the Pulmonary Team are available 24 hours a day by phone at (614) 722-4766 to answer questions about your child's health.

If you have any questions, be sure to ask your doctor or nurse.