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CYSTIC FIBROSIS

Cystic fibrosis (CF) is an inherited disease that affects the digestive system and the glands in the lungs that produce mucus (Picture 1). It is not contagious ("catching"). CF also affects the glands that produce sweat and saliva. Glands are organs of the body that make important fluids such as mucus, saliva, and sweat.

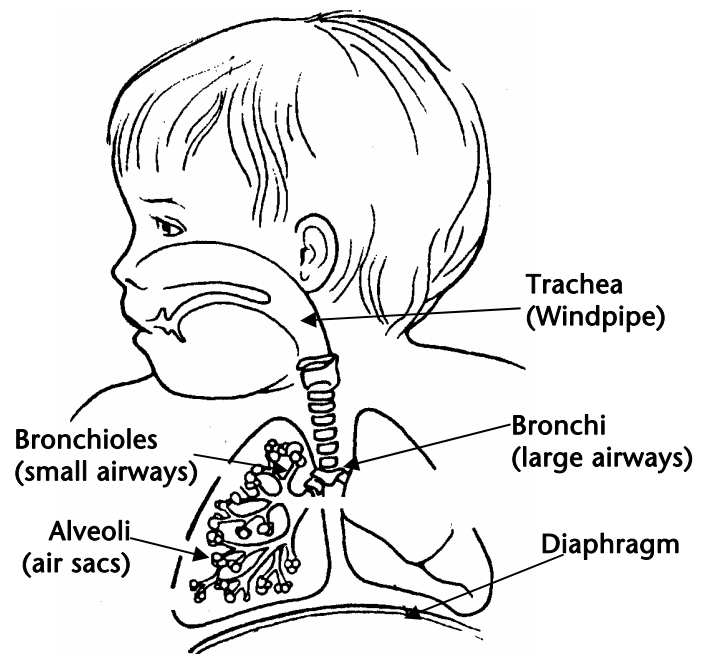
WHAT CAUSES CYSTIC FIBROSIS

Chromosomes (CROW-ma-somes) are in every cell of the body. The chromosomes carry the genes which decide hereditary traits. 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 genetics of CF in more detail are available from the Cystic Fibrosis Foundation. A genetics counselor can also be very helpful to families of 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, or weight loss
- Cough that does not go away or comes back often
- Wheezing
- Large, loose frequent stools, oily stools, or frothy and foul-smelling stools
- Chronic sinus infections

A child who has any of these symptoms should be seen by a doctor. Tests can be done to find out if CF is the cause or if some other condition is present.



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 stool 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 collecting a sample of blood and testing it.

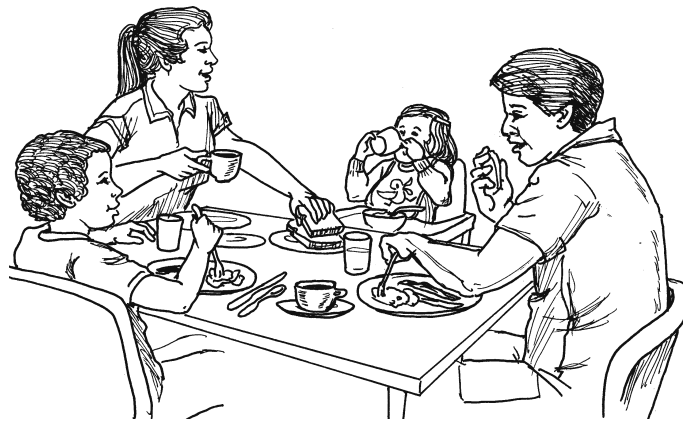
CARE OF THE LUNGS

- One of the most important things you can do to care for your child is to keep the airways of the lungs free of thick mucus. If mucus collects in the lungs, germs can grow in it and cause lung infections.
- Aerosols may be needed to keep the airways moist and the mucus thin. A nurse will instruct you and your child on how to give aerosol treatments.
- Antibiotics are often prescribed for even mild "cold like" symptoms.
- No one should smoke around your child.
- Sometimes the child may need to be hospitalized and receive IV antibiotic therapy to help keep the 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.
- There are other methods of giving CPT that are used. :Your child's nurse, doctor, or respiratory therapist will help decide which treatment would be the most effective for your child.
- It is **very important** to do **all** of the treatments!

NUTRITION

Your doctor and dietitian will help you plan your child's 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 stools.
- Your child may need to eat more often. Three well-balanced meals and 2 to 3 between-meal snacks are usually recommended (Picture 2). **Do not let your child eat all day long or snack continuously throughout the day.**
- Enzymes (EN-zimes) are found in the digestive juices. These substances help digest food. With CF, enzymes made by the pancreas are sometimes blocked and can't reach the small intestine. If this happens, capsules (containing tiny beads) must be taken with meals to help digest food. When enzymes are added to the diet, they help to maintain good nutrition and growth. Enzymes also help the child to have normal bowel movements.
- Your child will need salt added to the diet to replace the salt that is lost in the 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.
- Vitamin supplements are often needed. Your child's doctor will order the diet enzymes and vitamins your child needs.
- A dietitian from the Pulmonary Team will spend time talking with you about the foods which will be best for your child.



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

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 all booster shots are given.
- 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 the child's development.
- Your child will learn to pace himself and not become too tired from play.
- 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 the child has CF (Ask your nurse for another copy of this Helping Hand). The teacher and other students should be told that CF is **not** contagious. If medicines are to be taken 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. Good communication with the school is important.

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.