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Cystic Fibrosis (CF)

What is cystic fibrosis?

Cystic fibrosis (say:SIS-tik fie-BRO-sis) or CF is a disease that mainly affects the lungs and digestive system. Although CF can be treated and CF patients can usually lead fairly normal lives, there is no cure for CF. With good medical care the majority of children is surviving to adulthood.

CF affects the lungs

Normal mucus is thin and slippery. It keeps the lungs clean by removing dirt and germs. In CF, mucus is sticky and clogs the tubes of the lungs. This makes it hard to breathe. Bacteria then collect in the tubes. This leads to cycles of infection and swelling. These cycles damage the lung tissues.

CF may also affect the digestive system

CF may also affect the digestive system. Mucus blocks the ducts (tube-like channels that carry fluid) of the pancreas. The pancreas is an organ just below the stomach that makes enzymes to help digest food in the small intestine. Enzymes are proteins in cells that speed up reactions. Many enzymes help with digestion.

When the ducts from the pancreas to the small intestine are blocked by mucus, the enzymes cannot reach the small intestine. This means food is not properly digested. When this happens, a child with CF does not get enough nutrition from food.

CF is a genetic disease

Children are born with CF. CF is a genetic disease, meaning it is passed from parents to their children. About 1 in every 25 people carry the gene that can cause CF. A gene is a section of DNA that gives an instruction to a cell. Most of the time, the instruction is a “recipe” for making a protein.

CF is caused by a recessive gene. This means that a person needs to have 2 copies of the gene to develop CF. If a person has only 1 copy of the gene, they will not have CF, but they may pass the gene on to their children. People with only 1 copy of a recessive gene are called “carriers” of the gene.

Most parents do not know they carry the CF gene because they have only 1 copy, so they do not have symptoms. To develop CF, a child must inherit 2 copies of the gene, 1 from each parent.

Two parents with the CF gene may have many children with CF or none at all. The risk of having a child with CF is the same with each pregnancy.

CF is not contagious. You cannot catch it from someone else.

Signs and symptoms of CF include:

- Recurrent chest infections.
- Constant cough that expels thick mucus.
- Having a very big appetite with weight loss.
- Bowel movements that are bulky, frequent, and foul-smelling.
- Skin that tastes salty.
- Failure to grow or gain weight.
- Meconium ileus: a blockage in the small intestine by the newborn’s feces.
- The symptoms of CF are often confused with other conditions. For example, asthma, chronic bronchitis or pneumonia, and celiac disease can have similar symptoms as CF.

How CF is diagnosed

Doctors usually order a sweat test if they suspect CF. This is a simple test that measures the amount of salt in the sweat. It will not hurt your child. Heat or medicine is applied to a local (small) area of the skin. If the sweat contains more salt than usual, it points to CF.

- A test to check for enzymes in the intestine may be performed.
- Chest X-rays may be taken to see if there are any changes in the lungs.
- More recently, genetic tests are used to diagnose CF. Genetic tests can diagnose CF before a child is born.

How CF is treated

There is no cure for CF. With adequate treatment and regular follow-up, most children with CF can live fairly normal lives.

CF treatment is tailored to the child’s needs. It also depends upon the stage of the disease and which organs are affected.

Treating the lungs

Most CF treatments focus on the lungs. The treatments work to loosen and thin the mucus that clogs the airways.

Treatments for the digestive tract include:

- Taking pancreatic enzymes with meals to help digestion.
- Taking supplements and vitamins to promote good nutrition.
- Eating a special diet with increased calories and protein.
- Adding salt to the diet to replace the excess amounts lost during sweating.

Activity

Children with CF should be allowed to play games and sports. Sports such as running and swimming are often helpful because they clear the lungs of mucus.

Children with CF loose more salt during exercise and hot weather than people without CF do. It is important to make sure your child replaces fluids and salt by drinking enough and eating enough salt.

Key points

- CF is a genetic disease that affects the lungs and digestive system.
- It is not contagious.
- With treatment and regular follow-up, most children with CF can live fairly normal lives and into adulthood.

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