CARING FOR YOUR CHILD WITH: CYSTIC FIBROSIS

What Is Cystic Fibrosis?

Cystic fibrosis (CF) is a lifelong illness affecting cells that make sweat and mucus. Mucus is a slippery, somewhat sticky fluid that lubricates and protects mucous membranes. CF mucus is abnormally thick and sticky; it clogs the lungs and causes frequent lung infections.

CF also affects the pancreas, which has trouble making special chemicals called enzymes to digest food. Without enzymes, nutrients can't be absorbed from food.

CF is the most common fatal genetic disease in Caucasians in the United States (1 of every 2000 births). People with CF have a shorter life span, but with modern treatments, more people with CF live to middle age or older.

What Causes CF?

CF results from abnormal movement of the chemicals sodium and chloride (salt) in certain cells in mucous and sweat glands.

CF is inherited. A child must inherit a CF gene from each parent (called an autosomal recessive disorder). Both parents can be healthy but are carriers of CF. More than 12 million Americans carry the CF gene but have no symptoms.

What Are the Symptoms of CF?

Children with CF have normal lungs at birth, and symptoms may not appear until later.

Common symptoms are chronic cough and diarrhea, frequent bronchitis and pneumonia, bulky foul-smelling bowel movements (stool), large appetite in spite of poor weight gain, salty skin, and shortness of breath.

The first sign of CF in newborns may be a block of the intestines caused by increased thickness of the first stool.

How Is CF Diagnosed?

The health care provider will do a sweat test called pilocarpine iontophoresis. This painless test measures the amount of salt in the child's sweat. A high level can mean CF.

The health care provider can do genetic blood tests to look for changes inside cells. The health care provider can also do tests on bowel movements (stool) and blood to check effects on the pancreas, and get chest x-rays or do breathing tests to check the lungs.

CF can also be diagnosed with prenatal (before birth) testing.

How Is CF Treated?

Medicines help thin the mucus and prevent lungs from clogging. Antibiotics are also often given for infections. Enzyme supplements supply missing pancreatic enzymes. A special high-protein, low-fat diet may also improve nutrition.

Respiratory therapy is for the lungs. Thick mucus can be removed from lungs by tapping on the chest (chest percussion). Lying with the head lower than the feet can also help drain mucus. Lung transplantation is a treatment option in some cases and should be discussed with your health care provider.

DOs and DON'Ts in Managing CF

- ✔ **DO** follow your health care provider's instructions and give medicines and treatments as directed. Take your child to the health care provider at least three or four times yearly.
- ✔ **DO** avoid contact with people with respiratory infections.

✔ **DO** get your child a yearly flu shot.

✔ **DO** have your child drink plenty of fluids.

✔ **DO** have your child avoid gas fumes and smoke. These things irritate lungs and make CF worse.

✔ **DO** reach out to support groups.

⊗ **DON'T** neglect respiratory therapy, if advised by your health care provider.

⊗ **DON'T** give your child soy protein, which is hard to digest.

⊗ **DON'T** miss health care provider appointments.

⊗ **DON'T** stop prescribed antibiotics early or forget to do chest physical therapy.

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FOR MORE INFORMATION

Contact the following source:

- Cystic Fibrosis Foundation: Tel: (800) 344-4823; Website: http://www.cff.org

- American Lung Association: Website: http://www.lungusa.org
- National Heart, Lung, and Blood Institute: Website: http://www.nhlbi.nih.gov