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Related Articles published in the same issue	Mutation in the Gene Responsible for Cystic Fibrosis and Predisposition to Chronic Rhinosinusitis in the General Population XinJing Wang et al. JAMA. 2000;284(14):1814.

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Cystic Fibrosis

Cystic fibrosis is an inherited disease that affects tissues that produce mucus secretions. These include the tissues that line the airways in the lungs, the gastrointestinal tract, the ducts of the pancreas, and the ducts of the liver. Cystic fibrosis can also affect the sweat glands and the male reproductive system.

An article in the October 11, 2000,

issue of *JAMA* discusses a possible connection between the gene responsible for cystic fibrosis and chronic **rhinosinusitis**. Rhinosinusitis refers to inflammation of the mucous membranes that line the nose and facial sinuses. The **facial sinuses** are the cavities that surround the nose and are part of the upper respiratory tract.

WHAT IS CYSTIC FIBROSIS?

Cystic fibrosis is a disease that mainly affects the **respiratory system** (airways and the lungs) and the **digestive system** (stomach, intestines, colon, and other organs involved in the digestive process). Cystic fibrosis involves changes in the chemical properties of mucus, causing thicker-than-normal mucus that can lead to obstructions in the respiratory and digestive systems.

In the respiratory system, the abnormal mucus obstructs airways and also creates conditions that lead to repeated infections in the lungs.

In the digestive system, the abnormal mucus can obstruct ducts in organs involved in digestion. Cystic fibrosis can interfere with the normal processes of the liver, the **pancreas** (the organ that secretes the hormone insulin, which the body uses to break down sugar, and that produces digestive enzymes) as well as other organs that are part of the digestive process. This makes it difficult for persons with cystic fibrosis to effectively digest food and absorb nutrients.

WHAT ARE THE SIGNS AND SYMPTOMS OF CYSTIC FIBROSIS?

Since cystic fibrosis does not affect everyone in the same way, a person with cystic fibrosis may experience only some of the following symptoms.

An infant or child:

- May have a delay (more than 48 hours after birth) in the passing of **meconium** (their first stool)
- May have foul-smelling, pale, and greasy stools
- May lose weight (because of difficulty absorbing nutrients)
- May appear out of breath
- May have frequent wheezing
- May have a persistent cough that produces thick mucus
- May have frequent respiratory infections, such as pneumonia and bronchitis
- May have stunted growth (because of chronic malnutrition from difficulty absorbing nutrients)
- May have abnormally salty sweat
- May become easily dehydrated
- May have **nasal polyps** (fleshy growths inside the nose)

People with cystic fibrosis may also experience liver disease, diabetes, inflammation of the pancreas, and gallstones.

MANAGING THE DISEASE:

- Antibiotics for lung infections caused by bacteria
- Decongestants, **bronchodilators** (drugs that open airways congested with mucus), and anti-inflammatory drugs
- Chest or back clapping (to help loosen mucus from lungs) and postural drainage (to help drain mucus from lungs)
- Pancreatic enzymes (to help with digestion)
- A diet rich in proteins and calories
- Vitamins and other dietary supplements (to add more nutrients to the diet)

INHERITING CYSTIC FIBROSIS:

Cystic fibrosis is an inherited genetic disorder. A person inherits the disease from their parents. The gene that causes cystic fibrosis is a **recessive gene**, which means in order to get the disease you must inherit the gene from both of your parents. If you only inherit the gene from one parent, you become a carrier (you will not get the disease, but you can pass the gene on to your children). If you are a **carrier** and you and another person who is a carrier have a baby together, each child will have a 25% chance of having the disease and a 50% chance of being a carrier.

FOR MORE INFORMATION:

- National Heart, Lung, and Blood Institute
NHLBI Information Center
PO Box 30105
Bethesda, MD 20824-0105
301 592-8573 or www.nhlbi.nih.gov
- Cystic Fibrosis Foundation
800 FIGHT CF (800 344-4823) or
www.cff.org

INFORM YOURSELF:

To find this and previous *JAMA* Patient Pages, check out the AMA's Web site at www.ama-assn.org/consumer.htm.

Additional Sources: National Heart, Lung, and Blood Institute, National Institute of Diabetes and Digestive and Kidney Diseases, Cystic Fibrosis Foundation, The AMA Complete Guide to Your Children's Health, The AMA Encyclopedia of Medicine

Brian Pace, MA, Writer

Richard M. Glass, MD, Editor

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