



## Cystic Fibrosis

Cystic fibrosis is a genetic disease that causes the body to create thick mucus that builds up and obstructs ducts and tubes within the lungs, digestive tract, and pancreas. This build-up of mucus can cause severe and sometimes fatal infections, as well as malabsorption of nutrients, and it can also affect the sweat glands and male reproductive system.



Your doctor will likely perform blood tests and may test your sweat for high salt content, which can indicate cystic fibrosis. If the diagnosis is confirmed, your doctor may evaluate your condition with a chest x-ray, CT scan of the chest or abdomen, MRI of the chest or abdomen, abdominal ultrasound, or CT of the sinuses. There is currently no cure for cystic fibrosis, but there have been significant recent advances in the care of patients with cystic fibrosis that have significantly increased the expected life span over the past decades. Your doctor may recommend lifestyle changes, medications, chest therapies or surgery to help ease your symptoms.

## What is cystic fibrosis?

Cystic fibrosis is a life-threatening genetic disease that causes the body to create thick mucus. This thick mucus can build up and obstruct ducts and tubes within the lungs, digestive tract and pancreas. The build-up may cause severe and sometimes fatal infections and digestive issues. It may also affect the sweat glands and male reproductive system.

While cystic fibrosis may occur in people of any race, it is most common among Caucasians of Northern or Central European descent.

Symptoms of cystic fibrosis depend on the severity of the disease, but may include:

- Shortness of breath
- Frequent lung infections
- Persistent coughing
- Wheezing
- Fatigue

- Nasal congestion
- Stomach pain
- Weight loss
- Abnormal and foul-smelling stools
- Constipation
- Intestinal blockage
- Male infertility

There is now required screening for cystic fibrosis and other conditions at birth; therefore newborns are often diagnosed soon after birth, before symptoms occur. However, it is important to recognize early symptoms among infants, which include:

- Salty-tasting skin
- Lack of weight gain
- Delay in growth
- Rectal prolapse in which the rectum protrudes outside the body

## How is cystic fibrosis diagnosed and evaluated?

The most common types of cystic fibrosis testing include taking a blood sample for genetic testing or conducting a sweat test. A sweat test measures the amount of salt in a person's sweat. High salt presence can indicate cystic fibrosis.

The United States now requires screening of newborns for cystic fibrosis through testing blood samples for abnormal levels of pancreatic chemicals called immunoreactive trypsinogen, or IRT. In some cases, pregnant women can have their babies tested before birth through amniocentesis or a biopsy of the placenta, called chorionic villus sampling (CVS). Amniocentesis is the suction of fluid from the amniotic sac (the fluid around a developing embryo/fetus) through a needle inserted through the abdomen. This fluid can be then tested for cystic fibrosis. During CVS, a needle is used to remove a small amount of placenta, which is then evaluated for cystic fibrosis and other genetic diseases.

If you have been diagnosed with cystic fibrosis, your doctor may evaluate your condition using a stool test, spirometry or sputum culture. The following imaging tests also may be ordered:

- Chest or abdominal computed tomography (CT) scan: These exams use special x-ray equipment and computers to produce many detailed images of the inside of the lungs or intestines. These images can help determine the severity of cystic fibrosis by looking at the amount of mucus as well as looking for dilated airways in the lungs. The test can also look for infection. This test uses ionizing radiation. See the Safety page for more information about CT.
- Chest x-ray: This exam uses a small dose of ionizing radiation to produce images to evaluate for dilated airways containing mucus and also to evaluate lung infections that need to be treated with antibiotics. Chest x-rays are used periodically to observe changes in patients with cystic fibrosis and rule out other respiratory conditions such as pneumonia or a collapsed lung. See the Safety page for more information about x-rays.
- Chest or abdominal magnetic resonance imaging (MRI): These imaging exams use a powerful magnet, radio waves and a computer to produce detailed pictures of the lungs and digestive tract.

Although chest x-ray or CT is used more commonly for this disease, MRI can help assess damage caused by cystic fibrosis.

- **Abdominal ultrasound:** This imaging test uses a small probe, gel, and high-frequency sound waves, to produce pictures of the upper abdomen. It evaluates the pancreas, liver, and gallbladder, all of which are affected by cystic fibrosis.
- **CT of the sinuses:** This exam combines special x-ray equipment and a computer to produce multiple images of the paranasal sinus cavities. It can help identify unilateral nasal polyps, which are common in patients with cystic fibrosis.

## How is cystic fibrosis treated?

While there is no cure for cystic fibrosis, your doctor may recommend one or more of the following to help ease symptoms and improve quality of life:

- Lifestyle changes such as better nutrition, use of vitamins, increased physical activity, avoiding the use of tobacco and avoiding second hand smoke.
- Medications such as antibiotics and inhaled medicines can help open the airways and clear them of mucous and infection.
- Various chest therapies or airway clearance techniques (ACT) may help to loosen mucus, which makes it easier to cough up sputum and eliminate it from the lungs.
- In severe cases, surgery or other procedures may be needed to improve quality of life. These surgeries can include a lung transplant, procedures to stop bleeding from the lung, nasal polyp removal or the introduction of a feeding tube to help deliver proper nutrients.

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