

# What is Cystic Fibrosis

by: Christy Blackburn RN, ICU  
Candler County Hospital

*Cystic Fibrosis (CF)* is a chronic life-threatening disease that affects the lungs and digestive system. A defective gene causes the body to produce unusually thick, sticky mucus. This mucus clogs the lungs and can make breathing very difficult. It causes bacteria (germs) to get stuck in the airways that lead to severe lung infections which lead to lung damage. The mucus also blocks the digestive tract and pancreas which stops natural digestive enzymes from getting to the intestines to breakdown food for nutrients that the body needs to absorb in order to grow and stay healthy.

*Cystic Fibrosis* is a genetic disease which means people inherit it from their parent's genes (DNA). A person with CF has inherited two copies of the defective CF gene - one copy from each parent. If each parent is a carrier of the CF gene, their child has a 25% chance of having CF, a 50% chance of being a carrier and a 25% chance of not having CF or being a carrier.

The most common symptoms of CF are very salty tasting skin, persistent coughing, frequent lung infections, wheezing or shortness of breath, poor growth/weight gain in spite of a good appetite, and frequent greasy, bulky bowel movements.

A sweat test is the most common test used for diagnosis. A small electrode is placed on the skin to stimulate sweat glands. Sweat is collected and the chloride in the sweat is measured. A high level of chloride means that the person has cystic fibrosis. A genetic test is often used if the results from a sweat test are unclear.

Approximately 30,000 people in the United States have CF (70,000 nationwide) and about one in every 30 Americans carry the defective gene, but doesn't have the disease. *Cystic Fibrosis* is most common in Caucasians, but can affect all races. In 2006, the predicted median age of survival was 37 years. In 1955, children with CF were not expected to live even to the first grade. Today, people with CF are living longer healthier lives.

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Currently there is no cure for CF. However, specialized medical care, aggressive drug treatment and therapies along with proper CF nutrition can lengthen and improve quality of life. Treatment and therapies available for CF today vary with each person. Medication used to treat lung infections and help CF people breath better are mucus thinners, which are inhaled through nebulizers, antibiotics, anti-inflammatories and bronchodilators. Chest physical therapy is a technique used to help loosen mucus in the lungs so it can be removed by coughing. Percussion (hand clapping) or vibration over the chest wall loosens mucus. A vest may also be used. Deep breathing and huffing moves mucus into larger airways and stimulates coughing that removes the mucus. Supplements that help with food absorption/nutrition include pancreatic enzymes, multivitamins and high calorie/high protein drinks.

There are over 115 nationwide CF Foundation accredited care centers. These care centers work with each CF person individually to meet their specific needs and to keep them as healthy as possible. Today, thanks to continued research and specialized care, an increasing number of people with CF are living into adulthood and leading healthier lives that include careers, marriage and families of their own. For more information visit the CFF website at or phone 1-800-682-6858.