Definition
Cystic fibrosis (CF) is an inherited disease. It causes a defect in certain cells of the lung and digestive system. The defect makes the cells produce a thick, sticky mucus. This mucus can cause

Blockages in the lungs and airways

Problems digesting and absorbing nutrients

CF is a serious life-long condition, but the severity of the illness can vary greatly. The average life expectancy for someone with CF is about 35 years. Although, some with mild forms of CF can live to age 60 or beyond.

Causes
CF is a genetic disorder. A child with CF inherits defective genes from each parent. Parents who have the gene, but do not have CF, are called carriers.

Risk Factors
Factors that increase your chance of CF include:

Parents who are known carriers of the CF gene

Siblings with CF

Parents with CF—mostly the mother since men with CF are often sterile
What is Cystic Fibrosis? What Everyone Should Know

Symptoms
The abnormally thick mucus of CF blocks certain organs. This causes many of the symptoms of CF. Symptoms in infants may include:

- Difficulty passing the first stool
- Salty sweat
- Intestinal obstruction, sometimes requiring surgery

Mucus that causes blockages in the lungs may lead to:

- Coughing and wheezing
- Shortness of breath
- Difficulty with exercise
- Abnormally shaped clubbed finger tips
- Malformed chest

Mucus can also block the pancreas. This can block enzymes used to help you digest food. This can lead to:

- Bulky, bad-smelling, floating stools, due to poor digestion of fats
- Diarrhea
- Trouble gaining weight
- Poor growth

**Failure to thrive**

- Malnutrition
- Dehydration

Other symptoms may include:

- Jaundice or other symptoms of liver disease
- Chronic nasal congestion from chronic sinus infections

- Prolapsed rectum
- Nasal polyps

Excessive thirst or urination that may indicate diabetes mellitus type 2

- Stomach pain or swelling from intestinal blockage
Prevention of sperm production in males

Mildly decreased fertility in females

Overall, girls are affected more severely than boys.

**Diagnosis**

The doctor will ask about symptoms and medical history. A physical exam will be done. CF is suspected in a child with classic symptoms, especially if a sibling has CF.

CF is often diagnosed by symptoms, family history of CF, or a positive screening test in newborns. The diagnosis may be confirmed with genetic testing. Other lab tests that may be used to confirm CF include:

- Sweat chloride testing
- Transepithelial nasal potential difference measurement

Your doctor may need to check your lungs. This may be done to look for symptoms or to determine treatment. Tests may include:

- Chest and/or sinus x-rays
- Lung function tests
- Sputum cultures

Tests may also be needed to check the pancreas. These tests may be done to assess symptoms or determine treatment.

**Treatment**

There is no cure for CF. Treatment is aimed at:

- Improving the amount of nutrition your body receives
- Preventing and treating lung and sinus infections
- Keeping the airways and lungs as clear as possible

Treatment for CF includes:

**Nutritional Support**

Better nutrition will help improve overall health. It will also improve growth and development in children. Children who have returned to normal weight within 2 years of the diagnosis have fewer coughing episodes and better lung function. Some nutritional steps that may help include:

- A high-calorie diet planned by a registered dietitian
- Nutritional supplements, including fat-soluble vitamins
- Pancreatic enzyme tablets with meals to improve digestion and absorption of nutrients
- Drinking lots of fluids and salt replacement, especially in hot weather or during illnesses
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Managing Lung Infections
Thick mucus in the airways increases the risk of respiratory infections. The infection can also be more severe because of the mucus. Treatment of a current infection often requires antibiotics. Prevention of new infections may be done with:

- Vaccination
- Antibiotics (usually inhaled)

Lung and Airway Support
Medications help keep the airways clear. Most will be delivered through an inhaler or nebulizer. Medications may include:

- Bronchodilators—to relax muscles and open the airway
- Mucolytic agents—to reduce mucus and help it move out of the lung
- Steroid inhalers—to decrease swelling and irritation (only when necessary)

Other steps that may help clear mucus from the lungs include:

- Hypertonic saline is a special type of salt water. A nebulizer machine creates a mist of this saline, which is inhaled. The mist may help thin out the mucus in the lungs.
- Chest percussion is rhythmic clapping over the chest. They may help clear mucus from airways. It is most helpful if done at least twice per day.
- Oxygen therapy may be required as the disease progresses. Ventilation may also be needed. Talk to your doctor about whether these are options for you.

Other Treatment
Surgery may be required to treat blockages in the intestine. Lung and liver transplants may also be considered. Support is important for those with CF and their families. Ask you doctor about local support groups or counseling options.

Prevention
If you have the defective genes, there is no way to prevent CF. Adults can be tested to see if they carry the genes before having children. Prenatal testing can determine if a baby will have CF.

RESOURCES:
- American Lung Association
  http://www.lung.org
- Cystic Fibrosis Foundation
  http://www.cff.org

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Community/Support

- **CF2Chat:**
  A forum and chat website for people with cystic fibrosis. The site also includes a section called CF in the News, a photo gallery and a recipe page. There is also an extensive section dedicated to therapy and health tips.

- **CysticFibrosis.com — Forums:**
  The heavily trafficked forums section of CysticFibrosis.com includes user blogs. It boasts over 7,500 members and a searchable database of over 20,000 topics.

- **Cystic-L:**
  Cystic-L is a free email service dedicated to the exchange of information and support specific to cystic fibrosis. Members include those with cystic fibrosis, as well as family members, medical professionals, researchers, and more.

- **CysticLife Community:**
  CysticLife.org is a social network just for the cystic fibrosis community. This website provides a place for the CF community to share tips, questions, ideas, experiences and encouragement.

- **Facebook.com Group — Cystic Fibrosis:**
  A group page within the Facebook community network designed for people with cystic fibrosis, family members, and doctors who treat it. A place to share information, personal experiences and stories, and offer support and advice.

- **Facebook.com Group — Adults with Cystic Fibrosis — A living legacy!:**
  “A place where adults living with cystic fibrosis can share their life stories, adventures and goals. Coming together to support each other.” Includes many relevant discussion topics and many heartfelt posts from the over 200 group members.