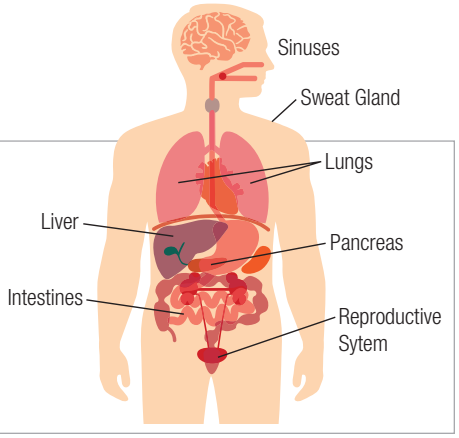
Twenty Facts About

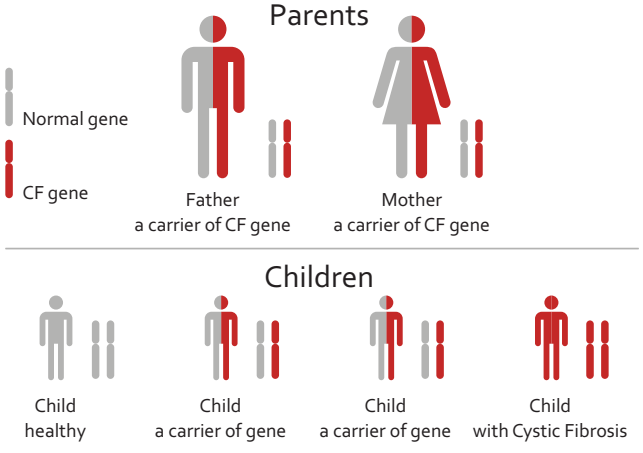
Cystic Fibrosis

Cystic Fibrosis is a progressive genetic disease that affects the lungs and other body systems and for which there is currently no cure. With new therapies, the life expectancy and control of CF disease continues to improve.



1. Cystic fibrosis (CF) is one of the most common genetic (inherited) diseases in North America.
2. CF occurs when a person inherits two mutated (abnormal) CFTR (cystic fibrosis transmembrane conductance regulator) genes, one from each parent. Approximately 2000 CFTR gene mutations have been linked to disease.
3. CF is inherited as an autosomal recessive disease, meaning only people with 2 CFTR mutations have the disease. People with only one CFTR mutation are carriers and do not have the disease. If both parents are carriers, there is a 1 in 4 chance that their child will receive an abnormal CFTR gene from each parent and have the disease.

Inheritance of Cystic Fibrosis



4. While CF is more common in Caucasians, CF occurs in all races and ethnicities.
5. While there is no cure, life expectancy has steadily improved with the median survival exceeding 45 years in the United States depending on when a person was born. Death is most often due to progressive lung disease.
6. There are now more adults than children with CF in the United States.
7. The defective gene causes the CFTR protein to not form or work properly, causing abnormal movement of salt and water across the cell, which leads to dehydration of the airway surface and thick mucus that clogs ducts throughout the body.
8. The sweat chloride measurement, or sweat test, is the recommended test to diagnose CF, since affected people have higher sodium and chloride levels, making the sweat more salty.
9. Newborn screening for CF done on blood samples can identify most children before one month of age, which allows for early treatment and disease monitoring.
10. Older children and adults are usually tested for CF because of symptoms such as frequent respiratory infections, malnutrition, and/or male infertility.

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11. New medications can now improve CFTR function in the cells of many patients based on the type of gene mutations they have.
12. People with CF have abnormally thick mucus, which blocks the airways (obstruction) and leads to repeated infections and damaging inflammation in the lungs. Treatments are directed at trying to prevent and treat these problems.
13. Infection control is important to help avoid exposure to dangerous pathogens (germs including bacteria and viruses that cause illness). People with CF are discouraged from close contact with other CF patients, and should try to limit exposure to certain bacteria and viruses.
14. Immunizations, including a yearly flu shot and COVID-19 shots if eligible, are important preventive measures and recommended for people with CF and their families.
15. Due to the thick mucus blocking the ducts in the pancreas and preventing the release of pancreatic enzymes that help break down fat and protein, most people with CF suffer from malabsorption of food, leading to unhealthy low weight and nutritional issues. In addition to needing to enzyme replacement, they need to take extra fat-soluble vitamins (A, D, E, and K) and calories to stay healthy.
16. The thick mucus also blocks ducts in other organs, causing additional health problems including liver disease, CF related diabetes and pancreatic insufficiency. Other health issues can develop in CF such as sinus disease, osteoporosis, and reproductive health issues. The CF healthcare team helps monitor and manage them.
17. CF care requires a team that includes the person's family, a primary care doctor, as well as CF doctors, nurses, dieticians, social workers, pharmacists, respiratory therapists, and others. There is a network of specialized care centers in the United States that are accredited by the Cystic Fibrosis Foundation (CFF) offering patients high quality care. To find a care center, contact the Cystic Fibrosis Foundation website (www.cff.org/Care-Centers/Find-a-CF-Care-Center).
18. Regular monitoring of lung function tests and sputum cultures helps the healthcare team advise individuals with CF on how to keep their lungs healthy and adjust treatments with any change. The CFF recommends that everyone with CF be seen in a specialized care center at least every 3 months.
19. Resources are available to help people with CF struggling with insurance, healthcare, school or other life or work issues. Contact the CFF Compass Program (www.cff.org), Cystic Fibrosis Research, Inc. (www.cfri.org), or ask your CF healthcare team for help.
20. Good adherence with home therapies as they are prescribed is a key to living as healthy and long a life as possible. People that have trouble doing their home treatment plan should talk to their healthcare team.

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Other Resources:

Cystic Fibrosis Foundation
www.cff.org

American Thoracic Society
www.thoracic.org/patients

Cystic Fibrosis Research, Inc.—an American Thoracic Society PAR organization
www.cfri.org

European Cystic Fibrosis Society (ECFS)
www.ECFS.org

Cystic Fibrosis Trust (United Kingdom)
www.cysticfibrosis.org.uk

Boomer Esiason Foundation
www.esiason.org

United States National Library of Medicine
www.medlineplus.gov/cysticfibrosis.html