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**Pulmonary Disorders 2017: Cystic fibrosis: A success story - Shahid Sheikh - Ohio State University College of Medicine, USA**

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Cystic fibrosis is an inherited illness which affects the lungs and digestive system. When mucus clogs the lungs, breathing can get really hard for a human. The thick mucus also allows germs to thrive and multiply, leading to infections and inflammation and often leading to severe damage to the lungs and respiratory failure. The buildup of mucus in the pancreas prevents digestive enzymes from being released which help the body break down food and absorb important nutrients. Cystic fibrosis in a gene called CFTR (cystic fibrosis transmembrane conductance regulator) is caused by a change, or mutation. This gene controls the salt and fluid flows within and out of your cells. If the CFTR gene is not working the way it should be, a sticky mucus is building up in your body. To get CF, both of your parents must inherit the mutated copy of the gene. Ninety percent of those affected by the F508del mutation have at least one copy. People with CF are often malnourished and growing poorly. The thick mucus can also block the liver bile duct, which can cause liver disease in some people with CF. CF may have an impact on men's desire to have children. CF has no impact on sexual growth in either men or women, however. Babies with CF are growing, evolving and doing what other infants do. CF children go to school, play sports and get driver's licenses. People with CF are allowed to go to college, pursue successful careers and have their own families. Today, more than half of people with CF are 18 years of age or older, due to improved medical treatments and care. Many people with CF can expect healthy living and fulfilling life in their 30s, 40s and beyond. A full CF diagnostic evaluation will include a sweat chloride test, a genetic or carrier test and a clinical examination at a CF Foundation-accredited treatment centre. Most children are now screened for CF by newborn screening at birth, and the majority are diagnosed with age 2. Some people with CF are however diagnosed as adults. A physician seeing CF symptoms will order a sweat test and a genetic test to confirm the diagnosis. People of all ages with CF have to follow a regular treatment routine to stay healthy. In addition to a diet and exercise program, there are medicines that help clear the dense, sticky mucus from the airways and minimize inflammation, antibiotics to treat CF lung infections and new therapies for imprinting the defective CFTR protein in CF.

The body develops thick and sticky mucus that can block the pancreas and clog the lungs. Cystic fibrosis (CF) can be life-threatening and people with the condition tend to have a life span that is shorter than normal. CF is a genetic disorder that primarily affects the lungs and digestive system, but it can lead to fatal complications such as hepatitis and diabetes. The faulty gene responsible for CF results in the production of thicker, more sticky mucus than normal. It's hard to cough the mucus out of the lungs. This can make it hard to breathe and lead to severe lung infections. The mucus also interferes with pancreatic function by preventing the proper breaking down of food by the enzymes. The result is digestive problems which could lead to malnutrition. This mucus thickening may also cause male infertility by blocking the deferens vas, or the tube which carries the sperm from the tests to the urethra. Lung obstruction by CF increases the risk of lung infections such as bronchitis and pneumonia because it creates optimal conditions for pathogens to grow. Pancreatic obstruction can lead to malnutrition, and poor growth. It was also linked to an increased risk of diabetes and osteoporosis. CF isn't currently being cured. However, medication can control the disease 's symptoms, and improve the quality of life. Symptoms can vary and plans for treatment will be individualised. There are alternate ways to treat CF which do not involve the airways. Implanted devices can allow the frequent and routine administration of drugs for long-term access to the bloodstream. They can make more effective and less intrusive management of a chronic condition such as CF.

CF is an inherited disease. For someone to have CF, both of their parents will inherit the defective gene. The

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defective gene contains codes that control the flow of salt and water outside the organs, including the lungs and pancreas, to produce a protein. In CF, the salt balance is disturbed which leads to too little salt and water outside the cells and production. CF screening is done to all newborns in the U.S. by testing a small blood sample or sample. This may indicate a baby may have a health condition and need further investigation. CF is normally diagnosed by a sweat test. Sweat is collected, and chloride, a salt component in the sweat, is measured. A high chloride level is a clue for CF. It is also possible to conduct genetic tests by testing cheek cells or a blood sample. These tests are primarily used to assess if a person carries the CF gene, but they can also be used to confirm a CF diagnosis after an ambiguous sweat test result.

Cystic fibrosis (CF) is an autosomal recessive progressive disease involving many organ systems but primary morbidity and mortality is with the involvement of lung disease. Because of genetic defects, inadequate hydration of pulmonary secretions leads to thick mucus causing airway obstruction which predispose to colonization, chronic infection and inflammation leading to irreversible bronchiectasis and ultimately resulting in end stage lung disease and respiratory failure. Current therapies for pulmonary disease in patients with CF decrease disease progression by improving secretion mobility and decreasing pulmonary infection and inflammation. These therapies have improved median predicted survival among patients with cystic fibrosis to more than 40 years. Now new disease modifying therapies are becoming a reality. We will discuss how life expectancy have improved over time and how it will be keep on getting better as CF now is not considered a pediatric disease. We will also discuss CF care model and its implications on other chronic illnesses.

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