What is Cystic Fibrosis?

Cystic Fibrosis, (CF) is an inherited genetic disease that affects a number of organs in the body, primarily the lungs and pancreas by clogging them with thick and sticky mucus. Repeated infections and blockages can cause irreversible lung damage and death. Mucus blocks the tiny ducts of the pancreas which supply enzymes required for digestion, and consequently food is not properly digested and nutritional value is lost in the process.

The sweat glands are also affected and the body may lose an excessive amount of salt during exercise or hot weather.

In early childhood, prominent symptoms include growth problems or frequent infections, especially of the lungs. As the disease progresses, frequent lung infections (pneumonia) often lead to problems breathing, lung damage, prolonged courses of antibiotics, and respiratory failure requiring support by a ventilator. CF can also lead to frequent sinus infections, diabetes mellitus, difficulty with digestion and infertility.

The most consistent aspect of therapy in cystic fibrosis is limiting and treating the lung damage caused by thick mucus and infection with the goal of maintaining quality of life. Intravenous, inhaled and oral antibiotics are used to treat chronic and acute infection.

There is no cure for CF and most individuals with cystic fibrosis die young: many in their 20s and 30s from lung failure. However, with the continuous introduction of many new treatments, the life expectancy of a person with CF is increasing to ages as high as 40 or 50 in some CF individuals.

What causes Cystic Fibrosis?