

What is Cystic Fibrosis?

Cystic Fibrosis (CF) is an inherited, chronic, and progressive disease usually diagnosed in childhood that affects exocrine (outward secreting) glands. It is one of the most common genetic diseases affecting 30,000 children and adults in the United States. It is characterized by the production of abnormally thick and sticky mucus by the affected glands, which causes chronic and recurrent respiratory infections and impaired digestion.

One in every 30 people in the United States carries the altered CF gene. People who carry the gene have no symptoms of CF and many do not know they have the CF gene. The most common CF gene defect is Delta F508, but there are more than 1,000 different versions (mutations) of the gene. Symptoms vary from person to person, depending to some degree on the CF gene they inherited.

What are Exocrine Glands and How Does CF Affect Them?

Normally exocrine glands make thin slippery secretions like sweat, mucus, tears, saliva, and digestive juices. These glands and their secretions help the body to function normally. In CF, the secretions from exocrine glands are thick, dry, and sticky. These secretions build up and clog passages in many of the body's organs, but primarily in the lungs and digestive tract. In the lungs, the mucus can lead to serious breathing problems and lung disease. In the digestive tract, the mucus can lead to malnutrition and problems with growth and development.

What are the Initial Symptoms of Cystic Fibrosis?

- ❑ Salty-tasting skin
- ❑ Poor weight gain (even with good appetite)
- ❑ Abnormal bowel movements (chronic diarrhea, large, frequent, greasy, foul smelling stools)
- ❑ Wheezing
- ❑ Cough
- ❑ Increased lung mucus
- ❑ Pneumonia or other lung infection (recurrent or frequent)

Cystic Fibrosis Clinic

Since CF is a complex disease, it requires a multidisciplinary team approach. The CF care team here at The Children's Hospital is directed by a pediatric pulmonologist, Dr. Dion Roberts. He sees patients in the Pediatric Sub-Specialty Clinic at the hospital as well as at his office. At the Sub-Specialty Clinic there is access to a gastroenterologist and endocrinologist on a rotating basis. They, along with your pediatrician, can help to take excellent care of your child. Other equally important team members are:

- Clinic Coordinator : organizes healthcare team
- CF Nurse: patient/family education

- Respiratory therapist: plans, teaches, and carries out respiratory care
- Dietician: assists with attaining proper nutrition
- Social Worker: Assists with emotional well-being, as well as, psychological and financial concerns

What to expect in CF Clinic

CF Clinic is held the first and third and fifth (if applicable) Thursdays every month starting at 12pm. It is held at the Pediatric Sub-Specialty Clinic located on the third floor of Providence Alaska Medical Center. The appointments typically last between two and three hours. The CF Foundation recommends that children be seen in CF clinic at least every three months even when your child is well.

Medical Director:

Dr. Roberts is a pediatric pulmonologist. This means he is a doctor who specializes in the study and treatment of children with diseases of the lung. Even though he is a pediatric pulmonologist he still follows most of our adult patients with cystic fibrosis. His office phone number is (907) 561-5440

Community Events

Twice a year the CF clinic holds a family education day. This is usually a four hour event that includes the latest updates as well as lunch. It is a great opportunity to meet other families and learn about the latest CF information. There is one in the spring and one in the fall.

'Great Strides' is a national walk benefiting the CF Foundation. In Alaska, we have two walks sites. The first is in Wasilla and the second in Anchorage. They are held on different weekends usually in late May and early June. It is another excellent way to meet families, enjoy a bar-b-que and raise money for an excellent cause. Visit www.cff.org/greatstrides



In Alaska we have another annual fundraiser called the Lost Lake Run. This usually takes places the third weekend in August. It is nearly 16 mile run/hike outside of Seward that raises money for the CF Foundation. More information can be found at www.lostlakerun.org

Clinical Trials

Our clinic is currently participating in several clinical trials. Clinical trials are on-going studies of new drugs and therapies in the treatment of CF. Generally, most of the clinical trials our clinic participates in are those in which the child is six years of age or older. This is because most studies want the participant to be able to do pulmonary function tests appropriately. Check out more information at www.cff.org/research.