Cystic Fibrosis

Common Names

Cystic fibrous, CF

Causes/ Etiology

A defective gene causes abnormally thick and sticky mucus. This mucus builds up in breathing passages of the lungs and pancreas. The pancreas is the organ used to break down and absorb food. Collection of mucus results in life long infections and digestive problems. This can also influences sweat glands and a man's reproductive system. The individual must inherit two defective cystic fibrous genes.

Incidence

It is the most common deadly inherited disorder affecting Caucasians in the United States. Cystic Fibrosis affects 30,000 children and adults in the United States and 70,000 worldwide. 1,000 new cases are diagnosed each year. Most children are diagnosed by age 2. Those diagnosed by age 18 have milder forms of cystic fibrous. In the 1950's few children with cystic fibrous were able to complete elementary school but with advancements in medical treatment life expectancy has improved to 35 years of age.

Symptoms

Newborns:

- delayed growth
- difficulty gaining weight
- no bowel movements in the first 24-48 hours of life
- salty tasting skin

Symptoms related to bowel function:

- severe constipation relating to belly pain
- increased gas bloating
- belly appears swollen
- Nausea and loss of appetite
- Stools that are pale, foul smelling, have mucus, or float
- Weight loss

Lungs and Sinuses:

- Coughing or increased mucus

- Fatigue
- Nasal congestion caused by nasal polyps
- Recurrent episodes of pneumonia
- Sinus pain or pressure

Developmental Outcomes

Most children are fairly healthy until they reach adulthood. They are able to participate in most activities and attend school. Many young adults with CF finish college or find employment. Lung disease eventually worsens to the point the person is eventually disabled.

Assessment Approaches

- Blood test available looking for variations in genes
- Immunoreactive Trypsinogen-standard newborn screening test
- Sweat chloride test –high salt level is sign of the disease
- Fecal fat test
- Chest x-ray or CT scan
- Lung Function test

Treatment

Early intervention and a comprehensive treatment plan

Treatment for lung problems-

- antibiotics for treatment
- inhaled medicines
- lung transplant
- oxygen therapy
- care and monitoring at home
- special diet high in proteins and calories
- vibrating vests
- vitamin supplements

Care at Home

- Environmental factors at home such as dust particles, etc.

Role of the School Psychologist

- Children with cystic fibrosis will likely be in and out of the hospital because of the symptoms and infections that are a part of the disease. Therefore, school psychologists may play a role in easing this transition.
- Given the relatively short life expectancy of individuals with cystic fibrosis, the future may be daunting and a scary concept. School psychologist may help individuals process this idea and also help to address parent's concerns.
- School psychologists may also serve the role of educator to help teachers and students better understand the reason for his or her classmate's absence.

Resources for Parents, Teachers, and Professionals

Cystic Fibrosis Foundation
http://www.cff.org/

CysticFrobis.com
http://www.cysticfibrosis.com/

National Heart Lung and Blood Institute
http://www.nhlbi.nih.gov/health/dci/Diseases/cf/cf_what.html

Kids Health
http://kidshealth.org/parent/medical/lungs/cf.html