Cystic Fibrosis

Symptoms or Behaviors

- Persistent coughing (dry and hacky) and wheezing
- Excessive appetite but poor weight gain due to poor absorption of protein and fats
- Salty skin/sweat
- Easily fatigued
- Muscular weakness
- Noisy respiration
- Rounded shoulders, forward position of head, poor posture
- Enlarged fingertips
- Bulky, foul-smelling stools
- Repeated episodes of respiratory infections
- Decreased exercise tolerance
- Signs of emphysema

About the Disorder

Cystic Fibrosis is a chronic, genetic, progressive, non-contagious disorder of the respiratory and digestive systems characterized by the widespread presence of abnormally thick viscid mucus that clogs the lungs, interferes with breathing, and contributes to recurrent lung infections and progressive deterioration of lung function. It affects the pancreas, causing food to be poorly digested, and proteins and fats to be poorly absorbed. The sweat glands also produce an abnormally high content of salt.

CF cannot be cured; careful management and daily treatment will be required throughout life. Average life expectancy is mid-30's, with lung damage and heart failure frequently causing death.

It is the most common fatal genetic disease in the U.S. CF occurs in approximately one of every 2000 to 2500 births, equally in boys and girls, and one in twenty Americans is an unknowing symptomless carrier of the defective gene.

CF is generally diagnosed by determining salt content in chemically induced sweat.

The treatment depends on the stage of the disease and which organs are involved. One means of treatment, postural drainage (also called chest physical therapy – CPT) requires vigorous clapping and/or vibrating on the back and chest to dislodge the thick mucus from the lungs. Postural drainage, lying face down with head lower than feet will allow gravity to loosen secretions. Antibiotics are also used to treat lung infections and are administrated via pills and/or medicated vapors, which are inhaled to help open up clogged airways. Since the CF gene was discovered in 1989, the pace of CF research has greatly accelerated. Currently gene therapy offers the best hope for a life-saving treatment. This entails correcting CF cells by adding normal copies of the gene to repair the defective cells. Additionally, two other branches of research continue: protein and drug therapies; a cure for CF will likely need to combine all three. Drugs are being improved to thin mucus, reduce the number of respiratory infections, and improve lung function. The use of humidifiers and inhalers, as well as well as breathing exercises will also aid in loosening secretions.

Minnesota Low Incidence Project

Developed with Minnesota Low Incidence funds, Region 10 Physical Health Disabilities Network, and School Nurse Organization of Minnesota (SNOM)

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Educational Implications

Cystic Fibrosis does not affect a child's cognitive ability. Frequent and/or extended absences, however, may require re-teaching or special tutoring services. School personnel and peers should be inserviced about the special medical routines required for a student with CF, i.e. postural drainage, medications. The student him/herself may need support in dealing with the chronic illness, acceptance by peers, and self-image.

Instructional Strategies and Classroom Accommodations

- Curriculum modifications (extra time for assignments, every other problem, no time limits, alternative ways to test and get information, and resource room)
- Buddy system for notes, teacher outlines.
- Absences may require repeated instruction, modified requirements as noted above.
- Provide tutorial services/homebound instruction when necessary.
- Encourage the student to be as active and ambulatory as possible.
- Help the student to overcome the embarrassment caused by coughing and the need to expectorate mucus (make sure student has tissue and means of disposal at his/her desk).
- Allow the student to leave the classroom for a drink of water or have water at desk (may require more frequently than peers).
- Help student establish routine for taking medications and additional snacks.
- Provide a private room to complete medical procedures, if necessary.
- Allow a longer lunch time if needed.

Resources

Cystic Fibrosis Foundation (State)
1611 County Rd. B. West #221
St. Paul, MN 55113
(800) 344-4823
(651) 439-8283

Cystic Fibrosis Foundation (National)
6931 Arlington Road
Bethesda, MD 20814
(800) FIGHT CF
www.cff.org

Cystic Fibrosis: The Facts.
Harris, A. & Super, M. (1991)
Oxford University Press.

Taking Cystic Fibrosis to School, Special Kids in School, (2000). Jay Jo Books.

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