

What is it?

Cystic Fibrosis is a genetic condition involving the secretory glands that produce mucous and sweat. A child must inherit 2 copies of defective genes, one from each parent, to have the disease. A person is only considered a carrier if they have one copy of the defective gene. When 2 carriers have a child, the infant has a 25% chance of inheriting the disease.

Cystic fibrosis is a complex and unique disease. The mutated gene causes the body to build up thick layers of mucous that can affect the lungs, pancreas, liver, intestines and other organs. Mucous is a naturally occurring substance in the body that works to moisten certain organs and protect them from infection. But in children with CF, mucous becomes too thick and sticky to serve the same function.



What are the effects?

When mucous clogs the lungs, a person not only has trouble breathing but is also at greater risk for infection. Sticky mucous is a breeding ground to trap bacteria and allow it to multiply. Mucous also affects the pancreas by preventing the release of important digestive enzymes. Without these enzymes, the intestines can't absorb nutrients and break down food properly. CF patients often have bulky stools and experience gas, diarrhea, constipation and abdominal pain. Even in the liver, mucous can clog and block the bile ducts and therefore cause liver disease. CF also causes a person to lose large amounts of salt through sweat. Not only can this easily cause dehydration, but other medical concerns as well like fatigue, increased heart rate, and decreased blood pressure.

Symptoms and severity can vary from person to person. Lung function is often affected in early childhood years. Other times, symptoms do not present until teen to adult years. Common symptoms could include:

- Wheezing
- Salty skin
- Poor growth
- Low weight gain
- Persistent, productive cough
- Lung infections
- Shortness of breath
- Bulky stools

What is the treatment?

Respiratory failure is the leading cause of death in people with CF but advances today have dramatically increased the life expectancy of those living with the disease. *Early* and *consistent* treatment is imperative to the quality of life. Treatment plans are often individualized to each child but all will involve a combination of therapies.

- Airway clearance therapy: A routine to break up and loosen the mucous buildup. Vibrating vests are often worn by the child.
- Inhaled medication: Respiratory medications given via a nebulizer to help open the airways or thin the mucous. Antibiotics can also be prescribed.
- Pancreas enzymes: Medications taken with meals and most snacks to improve nutrient absorption.



Suggested school accommodations

Since children with CF are at increased risk for lung infections, they need to maintain a clean environment and limit their exposure to germs. Lung infections in people with CF do not pose an increased risk to the public. However, they do endanger others with CF. People with CF should maintain safe boundaries from each other and not share items.

- Extended lunch periods
- Snack breaks
- Flash pass for bathroom
- Flexibility of scheduling
- Modified workload, extra time on work
- Tutoring after absences
- Second set of books at home
- Fatigue considerations (consider locations of classrooms)
- Psychosocial considerations (like missed school, socialization)
- Preferential seating
- Adaptive PE consult
- Wipe down and clean desks
- Encourage hand washing
- Access to water or bottle to carry
- Do not discourage from coughing
- Tissues with trashcan near desk to dispose of



Exercise is a great way for children with CF to help clear their airways and strengthen their muscles. But remember, dehydration can become a concern because the loss of salt in their sweat (remember salty skin). Students should be encouraged to drink enough fluids and even avoid fluid-losing caffeine.

SHNIC school nurses information:

Specific health issues for individual health care plans

- Current medication orders for home and school, including vitamins
- Nutrition orders and fluid intake goals (high in protein, fat, salt and calories)
- Tube feeding orders and tube replacement per county policy, if applicable
- Plan for monitoring compliance of enzymes
- Exercise recommendations, adaptive PE if ordered
- Heat intolerance guidelines
- Emergency plan for respiratory distress
- Chronic care considerations (for flu season or event of hospitalization)
- Rest breaks as needed
- 504 considerations as needed

Resources & Manuals

Cystic Fibrosis Foundation
<https://www.cff.org/>

American Lung Association
<http://www.lung.org/lung-disease/cystic-fibrosis/>

Cystic Fibrosis in the Classroom
<http://www.cfnz.org.nz/wp-content/uploads/cfintheclassroom-2013.pdf>

School and cystic fibrosis– A guide for teachers and parents
https://www.cysticfibrosis.org.uk/media/151230/FS%20-%20School%20and%20CF_v4_Apr_2013.pdf