CYSTIC FIBROSIS

What is it?

Cystic Fibrosis (CF) is a progressive genetic condition that mainly affects the lungs and the digestive system. CF is a complex chronic disease that causes breathing difficulties and complications with how the body digests food. Mucous is a naturally occurring substance in the body that works to moisten certain organs and protect them from infection. In children with CF, mucous becomes too thick and sticky to serve the same function. The mutated gene causes the body to build up thick layers of mucous that can affect the lungs, pancreas, liver, intestines and other organs.

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 or break down food properly. CF patients often have bulky stools and can experience gas, diarrhea, constipation and abdominal pain. Even in the liver, mucous can clog and block the bile ducts which 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.

A child must inherit 2 copies of defective genes, one from each parent, to develop the disease. A person is considered a carrier if they have only one copy of the defective gene. If both parents are carriers of the disease, the child has a 25% chance of inheriting the disease. While children with CF who have lung infections don't pose an increased risk to the public, they do however pose a risk to other children with CF. Children with CF should maintain safe boundaries from each other and not share personal items.

What are the symptoms?

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. The types and severity of symptoms can vary greatly from person to person diagnosed with CF. Often lung function is affected in early childhood years. Other times, symptoms do not present until teen to adult years. Common symptoms could include:

- Salty-tasting skin
- Persistent coughing
- Frequent lung infection, including bronchitis or pneumonia
- Shortness of breath, wheezing
- Poor growth or weight gain despite normal appetite
- Frequent greasy, bulky stools, constipation
- Male infertility

What is the treatment?

There is no known cure for CF but medical advances have dramatically improved the life expectancy for those living with the disease. Early and consistent treatment is imperative to improving quality of life. Treatment plans are individualized to each child but all involve a combination of therapies. Airway clearance therapy is routine to break up and loosen the mucous buildup. Vibrating vests are often worn by the child. Respiratory medications given via a nebulizer are also administered to help open the airways or thin mucous secretions. Antibiotics can also be prescribed to fight bacterial infections. Pancreatic enzymes aid in food digestion and nutrient absorption. Enzymes are taken with meals and most snacks.

Kennedy Krieger Institute

The **Specialized Health Needs Interagency Collaboration (SHNIC)** program is a collaborative partnership between the Kennedy Krieger Institute and the Maryland State Department of Education.

Suggested school accommodations

CF does not affect a student's cognitive ability. A student with CF may have respiratory and digestive problems that range from mild to severe; therefore possibly requiring special accommodations in a school setting. In order to decrease their risk, the student must maintain a clean environment and limit their exposure to germs. Supporting students with this condition in the school require educators and parents/guardian to work as a team. Some accommodations to consider for a 504 could include:

- Extended lunch periods
- Snack breaks
- Flash pass for bathroom
- Adaptive PE consult
- Flexible scheduling
- Modified workload, extra time on work
- Extra set of books at home
- Consider location of classroom and school layout
- Psychosocial considerations (like missed school, socialization)
- Consider wrap around service for missed school work and appointments
- Monitor for fatigue
- Wipe down and clean desks
- Encourage hand washing
- Allow student to keep hand sanitizer on desk
- Access to water or bottle to carry
- Do not discourage from coughing
- Tissues with trashcan near desk for disposal
- Maintain safe distancing from those who are ill (generally 6 feet is recommended)
- Temperature controlled environment including transportation
- Staff education/training as appropriate
- Emergency Evacuation Plan (EEP)

Specific health issues for Individualized Healthcare Plan

- 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
- Heat intolerance parameters
- Chronic care considerations (for flu season or event of hospitalization)
- Rest periods
- Communicate with school staff, parents, and provider any changes or concerns about the disease
- Emergency Care Plan(s) (ECP) related to medical needs in the school setting and staff education/training as appropriate for each

Resources & Manuals

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

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

Cystic Fibrosis Our Focus, School and cystic fibrosis: A guide for teachers and parents
http://cfnz.org.nz/wp-content/uploads/FS-School-and-CF_v4_Apr_2013.pdf

Cystic Fibrosis NZ: Starting school: A guide to CF for primary schools and teachers
https://www.cfnz.org.nz/assets/Guides-Fact-Sheets/4694137106/Starting-School-A-Guide-for-NZ-Primary-Schools-and-Teachers-FINAL.pdf