

CYSTIC FIBROSIS

Background

Cystic fibrosis (CF) is a progressive genetic condition affecting the respiratory and digestive systems. CF affects the cells that produce mucus, sweat and digestive juices. A mutated gene causes the body to produce abnormally thick and sticky mucous that can clog and damage the lungs, pancreas, liver, intestines, and other organs. Sticky mucous can trap bacteria leading to an increased risk of infection and prevent the release of important digestive enzymes. Without these enzymes, the intestines can't absorb nutrients or properly break down food.

Symptoms and severity can vary from person to person. Symptoms often do not present until teen to adult years, but lung function is affected in early childhood. Common symptoms could include failure to thrive or inability to gain weight despite caloric intake, persistent coughing, shortness of breath, wheezing, and frequent sinus infections. Children with CF 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.

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.

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. Airway clearance therapy is routine to break up and loosen the mucous buildup. Vibrating vests are often worn by the child during treatment. Respiratory medications given via a nebulizer are also administered to help open the airways or thin mucous secretions. Antibiotics can be prescribed to fight bacterial infections. Pancreatic enzymes aid in food digestion and nutrient absorption and are taken with meals and most snacks.



Top Takeaways for School Considerations

A student with CF may have respiratory and digestive conditions that range from mild to severe. CF does not affect a student's cognitive ability.

Students with CF are vulnerable to serious lung infections. School staff should support infection prevention guidelines to help minimize exposure to germs and bacteria including encouraging frequent hand hygiene and maintaining a safe distance from others who are sick.

If more than one student is diagnosed with CF in the school, it is important to implement safe physical boundaries. While separate classrooms can often be arranged, shared spaces like the cafeteria, nurses' office, and transportation may be more difficult. The rule of thumb is that children with CF should be separated by at least 6 feet.

Students with CF will need a special high fat and high calorie diet that includes pancreatic enzymes. Access to fluids and snacks may be necessary. A bathroom flash pass should also be considered.

Encourage the student to cover their coughs or sneezes with a tissue, dispose of properly, and perform hand hygiene. A trash can at their desk may be helpful.

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnosis of ineffective airway clearance, imbalanced nutrition: less than body requirements and risk for infection
- Current diagnosed health condition including date of diagnosis, progress of disease process and other chronic health conditions
- Current medication and treatment orders (consider schedule, equipment needs and side effects)
- Respiratory interventions and equipment needs
- Nutrition interventions and equipment needs (consider brand/size of feeding tube, tube replacement, water flushes, fluid intake goal and supplements); note school district policy on tube replacement and consider keeping backup feeding tube kit at school if applicable
- Temperature regulation considerations in school setting and transportation
- Activity, positioning, transferring (consider precautions and/or restrictions)
- Equipment troubleshooting (consider equipment/device user manual, battery, charger)
- Consider emergency care plan(s) (ECP) and emergency evacuation plan(s) (EEP) as related to medical needs in the school setting, and staff education/training, as appropriate

Discussion Starters for Educational Team

1. Would the student benefit from evaluations or assessments in any of the following areas: physical therapy, occupational therapy, speech and language therapy, assistive technology, adapted physical education, functional behavior, psychology, hearing and vision?
2. Would the student benefit from additional academic support and/or modified education (e.g., copies of notes, extra time, reduced workload, simplified instructions, alternative formats for presentation of material, 504/IEP)?
3. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule?
4. Does the student require activity precautions to prevent injury?
5. Does the classroom environment support the student's needs and/or equipment (e.g., desk/seating options, flash pass for bathroom or nurse)?

Resources

Cystic Fibrosis Foundation
cff.org

Starting school: A guide to CF for primary schools and teachers
cfnz.org.nz/assets/guidefactsheet



Scan QR code or visit KennedyKrieger.org/Redirect for more information.