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 gene mutation causes the body to produce abnormally thick and sticky mucus that can clog and damage the lungs, pancreas, liver, intestines, and other organs. Thick, sticky mucus can trap bacteria, increasing the risk of infection. It can also block the release of digestive enzymes, making it difficult for the intestines to properly break down food and absorb nutrients.

Symptoms and severity differ for each individual. Although symptoms may not become noticeable until the teenage or adult years, lung function is impacted early in childhood. Common symptoms may include:

- Persistent cough
- Shortness of breath, wheezing
- Frequent sinus infections
- Difficulty gaining weight despite caloric intake
- Failure to thrive

Children with CF often have bulky stools and can experience gas, diarrhea, constipation, and abdominal pain. Even in the liver, mucus can clog and block the bile ducts which can lead to liver disease. CF also causes a person to lose large amounts of salt through sweat. This can lead to dehydration, as well as other medical concerns such as 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 a safe distance 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 mucus 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 mucus secretions. Antibiotics may 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

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 the student's desk may be helpful.

Considerations for the Individualized Healthcare Plan (IHP)

- Nursing diagnoses: Ineffective airway clearance, imbalanced nutrition: less than body requirements, and risk for infection
- Respiratory interventions and equipment
- Nutrition interventions and equipment (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 action plans (EAPs) and emergency evacuation plans (EEPs) related to special health care needs, including staff education/training

Discussion Starters for the 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 strategies be implemented to assist the student with executive function (e.g., plan, prompts, organizers, agendas?)
4. Can rest breaks, safe spaces or reduced stimulation times be built into the student's schedule?
5. Does the student require activity precautions to prevent injury?
6. Will staff receive education/training to implement the student-specific emergency plan?

Resources

Cystic Fibrosis Foundation
cff.org

Starting school: A guide to cystic fibrosis for primary schools and teachers
cfnz.org.nz/assetsguidefactsheet

For more information, please scan the QR code or visit: KennedyKrieger.org/SHNIC

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