Cystic Fibrosis (CF)

What is cystic fibrosis (CF)?

- Cystic fibrosis (CF) is an inherited condition that causes mucus and secretions to become thick and sticky. These secretions can then block the lungs, gastrointestinal tract, sinuses, and other parts of the body.
- Children with CF are prone to recurrent pneumonia and to having digestive problems caused by the mucus blockage of the lungs and digestive system.
- As with most conditions, some children are more severely affected than others.
- Cystic fibrosis does not affect a child's intelligence or ability to learn.

How common is it?

Currently, about 30,000 children, adolescents, and young adults in the United States have CF.

What are some common characteristics of children who have CF or of CF as children present with it?

- These children may not appear ill at all, or they may be thin or small.
- They may cough frequently and may produce thick mucus. This cough is not contagious like the cough associated with a common cold or flu (ie, influenza); it is just the body's way of trying to clear mucus.
- Children with CF may have fingertips that are rounded or that have a blue tinge.
- Children with CF may require medications and lung treatments, but these medications will not affect attention or ability to learn.
- Some children with CF may have bulky stools or diarrhea and may pass gas more frequently.
- They may need to use the restroom frequently.
- Their skin may taste salty.
- Children with CF need to take pancreatic enzyme substitutes before eating, and they may need extra vitamins.
- Children with CF may have trouble gaining weight and therefore need to eat frequent high-calorie snacks.
- Many places have specialized CF centers; children should receive care from a specialist with expertise in this disease as well as from their pediatricians/primary care providers in the medical home.
- Ask the child's parents/guardians who is most involved in their child's care.

Who might be on the treatment team?

Children with CF are often cared for in specialized CF centers in addition to their medical homes. These centers will often include pediatric pulmonologists, respiratory therapists, nutritionists, and social workers with specific expertise in CF. Other specialists may include gastroenterologists.

What adaptations may be needed?

As always, the Care Plan should be updated after hospitalizations, emergency visits, child absences for illness, and changes in medications.

Medications

- Children with CF are often treated with oral and inhaled antibiotics.
- Nebulized treatments with medications such as albuterol may be given to open the airway.
- Other breathing treatments to loosen the mucus are also sometimes given.
- Pancreatic enzyme substitutes are needed to replace missing enzymes that the pancreas cannot secrete properly.
  - Store at room temperature in a safe location.
  - Consider allowing older children to carry the day's supply of enzyme substitutes themselves, so they do not have to spend most of their mealtimes waiting to receive medications, instead of eating.
  - Check the expiration date, and develop a system to obtain new supplies as needed.
- Vitamin supplements are usually needed.
- Special vaccines may be necessary for a child with CF. All children and staff should be fully immunized, including with influenza vaccine, to protect the child with CF.
- All staff who will be administering medication should have medication administration training (see Chapter 6).

Dietary Considerations

- Children with CF should have access to fluids throughout the day.
- Children with CF may need a little extra salt in their diets, especially when exercising in hot weather. They may also need a little extra fat and increased calories.
- Children with CF need to take enzyme substitutes before eating. For young children, the enzyme substitutes should be opened and sprinkled onto a small amount of food.

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- Older children can usually swallow capsules directly.
- Ask parents/guardians what foods are best to mix with the enzyme substitutes (applesauce is commonly used).

Physical Environment and Other Considerations

- Special equipment should be considered; some children use a special vest that vibrates to loosen mucus for chest physical therapy.
- Teachers and caregivers may schedule a visit with parents/guardians to review the specifics of the child's condition. The child may wish to briefly visit the child care center or school before coming for a full day, especially after hospitalizations.
- Open bathroom access is important.
- Children with CF should clean their hands frequently, and staff should be extra careful about cleaning toys and other children's hands.
- Chest physical therapy helps clear the mucus out of the airway; children with CF may need chest physical therapy while in child care.
- Children with CF should avoid sharing beverages or foods with other children and other activities that increase the chance of acquiring infections, because a cold can turn into pneumonia more easily.
- Encourage exercise.
- Children with CF can do almost anything, but they may need rest, if they tire more easily.
- Absences may occur more often because of worsening of respiratory or digestive symptoms. Help children with CF keep updated with their schoolwork and projects.
- Assist with a hospital or homebound program, a Section 504 plan, or an Individualized Education Program for children with frequent absences or hospitalizations.
- All adults working with children with CF should be up-to-date on their vaccinations, including their annual influenza vaccinations. All the children around them should also be up-to-date.

What should be considered an emergency?

- Call emergency medical services (911) for
  - Respiratory distress
  - Sudden chest pain
  - Significant blood in sputum
  - Any other acute worsening of the child's medical condition
- Call parents/guardians for
  - Fever
  - Change in sputum color or blood streaking
  - Increased fatigue

What types of training or policies are advised?

- Recognizing respiratory emergencies
- Background about CF
- Medication administration

What are some resources?

- American Lung Association: www.lung.org, 1-800-LUNG-USA (1-800-586-4872)
- Cystic Fibrosis Foundation: www.cff.org, 1-800-344-4823
- Local CF centers whose staff may be able to hold training sessions or provide materials

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