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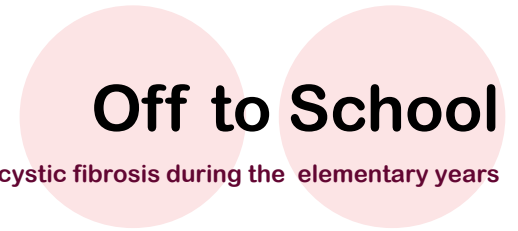
Together we can beat CF.

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Off to School

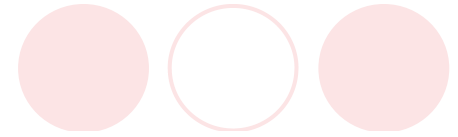
Managing cystic fibrosis during the elementary years





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Handling Difficult Situations

While cystic fibrosis (CF) is typically considered an “invisible” disease, some difficult situations teachers may encounter during the school year, as well as some tips on how to handle them, are listed below:

- 1) **Abdominal Pain:** If a student with CF complains of abdominal pain, the following interventions may be helpful: 1) pull the student aside and encourage him or her to visit the restroom to try and push out gas/stool, 2) have the child lie down, curl up, and apply a heat pad to the abdomen, and/or 3) give the child an analgesic like Ibuprofen or Tylenol. *If the child's abdominal pain is accompanied by fever or vomiting, or the pain is intense (student curls in fetal position) for more than 30 minutes, emergency care should be sought.*
- 2) **Coughing Fits:** Students with CF typically cough a lot, and may cough up large amounts of phlegm. For them, coughing is healthy and should never be discouraged. The cough is not contagious in any way. If the student has difficulty getting a cough under control, drinking water will help. *If the child starts coughing up large amounts of blood (cupfuls), emergency care should be sought.*
- 3) **Heat Stroke:** If a student with CF becomes dehydrated and overheated, the following interventions may be helpful: 1) move the student to a cool place, 2) loosen clothing, 3) apply a cold compress or fan lightly, and 4) push fluids. *If the child is unresponsive, has an altered mental state, or is vomiting, do not offer fluids and seek emergency care.*

Do's and Don'ts

- 1) **DO** encourage children with CF to cough as often as they need to. Coughing helps the child clear his or her airways.
- 2) **DON'T** draw attention to children with CF if they need to spit mucus into a tissue, step out of the classroom to get a drink to quiet their cough, or make an urgent trip to the restroom to eliminate stools. This part of the disease is very embarrassing for people with CF.
- 3) **DO** include children with CF in all physical activities. Physical activity helps clear the lungs, builds confidence, and makes the child feel like he or she is part of the group.
- 4) **DON'T** expose a child with CF to second-hand smoke, or have him or her clap erasers. Pollutants are harmful for weakened airways.
- 5) **DO** encourage children with CF (and their classmates) to wash their hands frequently, cover their coughs, and stay home if they are contagious. Viruses and colds can lead to pneumonias that may result in permanent lung damage.
- 6) **DON'T** treat children with CF differently. They want to fit in and feel “normal”; just like the other children in their class.
- 7) **DO** encourage children with CF to drink lots of fluid and eat salty snacks during warm weather or prolonged exertion. Children with CF sweat more than others, and consequently, are extremely prone to dehydration.
- 8) **DON'T** forget that the health of a child with CF is determined by what happens over the course of a lifetime; not what happens on a particular day. If a mistake is made with enzymes or a Vest treatment one day, the child's health will not be impacted.
- 9) **DO** encourage classmates to send notes and messages during a hospitalization. Hospital stays are difficult for a child with CF, and contact from “the outside” can lift his or her spirits.
- 10) **DON'T** be afraid to call the child's parents to report a decline in health, ask questions or report other concerns:

Students with Cystic Fibrosis

When reading this brochure, please bear in mind that cystic fibrosis (CF) affects each individual in different ways with varying degrees of severity. A person's health can change considerably from month to month — or even day to day. **There is no typical individual with CF.**

As a teacher, having a child with CF in your class means that you are teaching a child who has a chronic disease. Though it can cause severe respiratory and digestive problems, **CF has no effect on intelligence and is not contagious.** Because CF affects each person differently, the physical health and emotional attitude of a student with CF must be assessed on an individual basis. Whatever the age or condition, **always remember he or she is an individual, not a disease.**

Ana and Isabel Stenzel are identical twins with CF that survived double lung transplants. In their book, 'Power of Two', Isabel's post-transplant message about her identity was: "My CF was a huge part of my life, but a small part of who I was—beliefs, values, goals, or personality. I was a person, not just a diagnosis."



What is Cystic Fibrosis?

Cystic fibrosis (CF) is a life-threatening disease that causes mucus to build up and clog several organs in the body; particularly the lungs and pancreas. When mucus clogs the lungs, it can make breathing very difficult. The thick mucus also causes bacteria (or germs) to get stuck in the airways. **This leads to chronic inflammation (or swelling), lung infections, deadly pneumonias, and eventually, permanent lung damage.** In order to slow down the process of lung damage, people with CF inhale medications to open their airways daily, and take antibiotics to fight lung infections. In addition, daily airway clearance is performed to “shake” the mucus off the lung walls so it can be coughed up and out.

This tenacious mucus also blocks the digestive tract and pancreas. The mucus prevents digestive enzymes from getting to the intestines. **Without these digestive enzymes, the body is unable to break down food, grow normally, and remain healthy.** In order to thrive, people with CF need to replace these natural digestive enzymes by taking capsules filled with artificial enzymes. These capsules, taken with meals and snacks, help digest food and nourish the body.

Other parts of the body that may be affected by the oppressive mucus include, but are not limited to: the nasal passages (polyps, sinusitis), the stomach (acid reflux and cramping), the sweat ducts (dehydration), the intestines (diarrhea, constipation, gas pains and obstruction), and the reproductive system (sterility).

Psychological Health

A student with cystic fibrosis (CF) has the same emotional needs as others in your class. CF tends to make a child feel different. Even though the disorder can be categorized as “invisible” for the most part, it is not uncommon for a student with CF to be small in size, speak with a raspy or nasally voice, have a pale complexion, be barrel-chested, or develop clubbed fingers. **Group acceptance is an important need for these children.**

It may be helpful to have a classroom discussion about CF (if the student agrees). Peer teaching has been shown to be particularly successful. Thus, involving the parents and the affected child may be beneficial.

A child with CF may find it hard to participate in group situations. For example, shortness of breath may limit physical activity. A teacher can help strengthen a child’s self-image by encouraging the student to excel at what he/she does best, and by stimulating valuable relationships with other children by helping him/her gain acceptance in the classroom.

Finally, regular CF clinic visits, and possible periods of hospitalization, are facts of life for individuals with CF. During these absences, it is important that a child with CF keep pace with regular schoolwork. You may be asked to provide lesson plans and classroom materials to the parents. For the most part, clinic and hospital visits are pre-planned; giving you plenty of time to pull materials together.

Staying Healthy

Most people with cystic fibrosis (CF) can lead active lives—depending upon the severity of their disease—and slow the progression of their disease with airway clearance treatments, medication and regular visits to a CF Center.

Additional components to staying healthy include:

- 1) **Diet:** Since thick mucus gets in the way of proper digestion, enzyme supplements, vitamins, and a high-calorie, high-fat diet are vital for normal growth and development. Several studies show that nutrition and lung health are closely linked, so a proper CF diet is critical for optimal health.
- 2) **Germ:** CF puts the airways at risk for lung infections. One effective way to lessen the risk of developing infection is to limit contact with known germ sources. Although germs are everywhere and cannot be entirely avoided, a child with CF (and their classmates) should practice effective hand-washing with soap and water or hand gel, and avoid unnecessary contact with people who have a cold or any other contagious illness.
- 3) **Pollution:** Second-hand smoke from cigarettes, campfire smoke, chalkboard dust, and other forms of environmental pollution are particularly harmful to people with CF. In order to keep airways healthier, a child with CF should spend their time in a relatively “pollution-free” environment.
- 4) **Exercise:** Physical activity helps people with CF clear mucus from their lungs, builds confidence, and fosters inclusion. Children with CF should be included in all games and activities to the extent they are able to participate.

How Do People Get Cystic Fibrosis?

Cystic fibrosis (CF) is a genetic disease that people inherit from their parents through genes (DNA). **To have CF, a person must inherit two copies of the defective CF gene—one from each parent.** If both parents are carriers of the CF gene (i.e., they each have one copy of the defective gene but do not have CF), each child they conceive will have a 25% chance of having CF, a 50% chance of being a carrier like them, and a 25% chance of not having CF or being a carrier.

Approximately 30,000 people in the United States have CF. An additional ten million (or 1 out of 25 people) are unknowingly carriers of the defective CF gene. The disease is most common in Caucasians, but it can affect all races.

The severity of CF symptoms varies from person to person. The most common symptoms are:

- Very salty-tasting skin
- Persistent coughing, at times with phlegm
- Frequent lung infections, like pneumonia or bronchitis
- Wheezing or shortness of breath
- Poor growth/weight gain in spite of a good appetite
- Frequent greasy, bulky stools or difficulty in bowel movements
- Small, fleshy growths in the nose called nasal polyps

It is not uncommon for people with CF to be misdiagnosed with allergies, asthma, or chronic bronchitis.



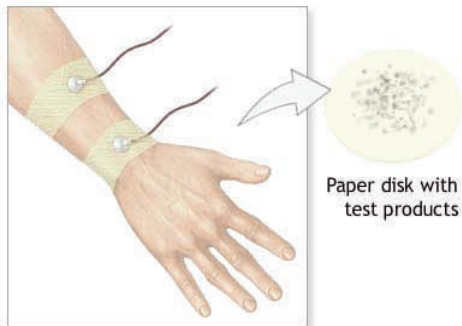
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How is Cystic Fibrosis Diagnosed?

Most people are diagnosed with cystic fibrosis (CF) at birth or before the age of 2. A doctor who sees the symptoms will order either a sweat test or a genetic test to confirm the diagnosis.

Since the basic problem with CF is an imbalance in the sodium chloride exchange in certain cells of the body, a sweat test is the most common test used to diagnose CF. A small electrode is placed on the skin (usually on the arm) to stimulate the sweat glands. Sweat is then collected and the amount of chloride, a component of salt found in the sweat, is measured. A high level of chloride—a score higher than 60 mmol/L—means that the person has CF.

In a genetic test, a blood sample is taken and sent to a laboratory to see if any of the various mutations of the CF gene are found. A genetic test is often used if the results from a sweat test are unclear.



Positive and negative electrodes applied

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The sweat electrolyte test is performed to determine the amount of chloride that is excreted in sweat from the body during a certain period of time. The test is used to determine if CF is present. Children with CF have increased levels of chloride in their sweat.

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Enzyme Dosing Chart

- 4) If a child forgets to take enzymes before a meal, they may be taken during or after the meal to help aid in digestion and eliminate some abdominal symptoms (do not give enzymes more than 30 minutes after the meal since they will not help beyond this point).
- 5) Children with CF must take in more calories, therefore they should be encouraged to finish meals and have as many snacks as they would like throughout the day.
- 6) Children with CF sweat more than their peers and have very salty sweat. Consequently, children with CF can lose a great deal of body salt through perspiration. **Encourage them to drink plenty of fluids and have a salty snack handy in warm weather or during prolonged exertion.**

Grams of fat/protein	Number of Enzymes	Examples
0	0	Water, juice, veggies, fruit snacks, hard candy
1-2	2	Fruit, pretzels, bread
3-4	4	Baked chips, crackers
5-6	7	Cookies, black olives, pancakes
6-10	8	Chips, low fat meats, noodles, candy bar
10-15	9	Nuts, cheese, high fat meats, PB sandwich
>15	10	Fast food, doughnuts, chicken skin

Taking Enzymes

Children with cystic fibrosis (CF) should be allowed to be responsible for keeping enzymes with them to avoid having them leave the classroom. There is no need to lock enzymes in a desk or store them in an office as they present no harm for others. In most cases, a student can carry a day's supply of enzymes in a lunch box and take them with meals and snacks.

If you will be supervising a child that needs to take enzymes, please note the following:

- 1) In general, this student has been prescribed _____ enzymes with snacks and _____ enzymes with meals. **The amount of fat, protein and starch in food varies greatly, therefore customizing the amount of enzymes based on the food being eaten is ideal.** The table on the next page may be used to help customize the enzyme dose. If you are not comfortable customizing the enzyme dose, simply give the child the dose prescribed above for snacks and meals.
- 2) **Enzymes must be taken immediately before eating because they only last 20 minutes.** If the student spends more than 20 minutes eating, an additional 1-2 enzymes should be taken.
- 3) **It is not dangerous for a child with CF to take too many or too few enzymes.** If the child takes too few enzymes (or completely forgets to take enzymes), he or she will experience abdominal pain, gas and distention until all of the undigested food is eliminated via stool.

Is Cystic Fibrosis Fatal?

Currently, there is no cure for cystic fibrosis (CF). However specialized medical care, aggressive drug treatment, chest therapy, and proper CF nutrition can lengthen and improve the quality of life for those with CF.

The best way for people with CF to fight their disease is to work with their medical caregivers at a CF Foundation-accredited care center. The care center partners with people who have CF to help keep them in the best health possible. To contact your student's CF doctor, please call:

There is no way to accurately predict how long people with CF will live, as many different factors may affect a person's health. Severity of disease and time of diagnosis are two such factors.

In the 1980's, children with CF rarely lived to age 15. **Today, more than _____% of all people with CF in the United States are 18 years or older, and the predicted median age of survival is _____years!**

People with CF are hospitalized frequently due to lung infection and scarring. Eventually, this scarring will lead to lung failure. If a lung transplantation is not successfully performed, lung failure will result in death.



What is a Typical Day Like?

While children with cystic fibrosis (CF) typically appear healthy on the outside, the disease takes its toll on the organ systems inside the body. **To slow the progression of the disease, the child with CF must follow a strict, time-consuming treatment regimen.** Airway clearance treatment is performed twice daily to clear mucus from the lungs. In addition, numerous medications are taken to combat malnourishment, lung infection, acid reflux, sinusitis, intestinal issues, and other problems that arise.

Having a chronic disease means that painful and embarrassing symptoms are expected. Coughing fits, abdominal pain, sinus pain, gas, and frequent, smelly stools are considered “normal.” Although children with CF have good days and bad days, their overall energy level is not as high as other children’s. This perpetual fatigue is generally due to: their compromised airways, malnourishment, and their inability to get a good nights rest (due to night-time coughing and side effects from the medications).



Children with CF perform a minimum of one hour of airway clearance treatment each day.

Digestion

The gastrointestinal problems associated with cystic fibrosis (CF) result in the inability to digest fats, proteins and starches. **In order to digest food, people with CF must take pills called enzymes.** In addition, people with CF must eat a special diet high in fat and calories (think *anti-American Heart Association diet!*). When teaching your class about nutrition, be sensitive to the child with CF who has a diet that may appear unhealthy by most nutritional standards.

Your student will need to take enzymes with any meal or snack that has fat, protein or starch. These enzymes help the body absorb nutrients from food and reduce: abdominal pain, distention, the number/bulk of stools, and the amount of gas. Enzymes are not habit-forming nor will they alter a student’s attitude or emotional behavior. **Enzymes are naturally produced by our body, therefore they would not cause harm if accidentally ingested by a child that does not have CF.**

Left: Children with CF amaze their classmates (and teachers) with their ability to take medication by the fistful. Right: This shows the amount of medication the 4-year old child to the left must take each day.





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Airway Clearance

Frequent clearing of the throat and “coughing spells” are normal and healthy for people with cystic fibrosis (CF). To instigate coughing and subsequently clear mucus out of the lungs, airway clearance is performed for thirty minutes at least 2 times each day. If the lungs become infected, airway clearance may be performed 3 or more times each day. If you are required to perform airway clearance, follow these steps:

- 1) Place the following medication(s) in the nebulizer cup:

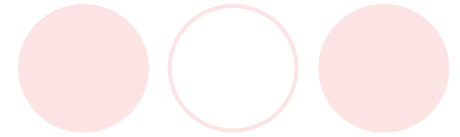
- 2) Fasten the jacket on the child and plug the tubes in tightly.
- 3) Press ON, program the display to 20/6/30 (from left to right), and press ON again to start the Vest.
- 4) Turn the nebulizer on, attach the tubing to the bottom of the nebulizer cup, hand the nebulizer to the child, instruct the child to breathe through the mouth (not nose).
- 5) Every five minutes, stop the Vest by pressing ON, reprogram using the chart below, remove the tubes, have the child cough twice to bring up mucus, plug the tubes in tightly, and press ON again to continue the treatment.

Hz	Freq	Time
20	6	30
19	6	25
18	6	20
10	10	15
9	10	10
8	10	5

6) Once smoke stops coming out of the nebulizer cup, remove the tubing and turn off the nebulizer machine (nebulizer typically finishes before the Vest).

7) After the Vest turns itself off (completion of treatment), remove the tubes and jacket, and have the child cough twice to bring up mucus.

Together we can beat CF.



Clinic Visits and Hospitalizations

People with cystic fibrosis (CF) visit a CF Center at least 4 times each year to monitor their health. During these lengthy visits, throat cultures are taken to identify which bacteria are present in their lungs, pulmonary function tests (PFTs) are performed to determine how well their lungs are functioning, and a physical exam is performed by a pulmonologist (lung doctor) to assess overall health and determine whether or not changes need to be made to medications. In addition, the following tests are performed at least annually to monitor all of the systems in the body that are affected by CF: a blood draw, x-rays, aerosol scans, bone density scans, and glucose-tolerance testing.

Even with all of this close monitoring and aggressive medical intervention, hospitalizations are an fact of life for people with CF. Typically, people with CF are hospitalized for routine “tune-ups.” These hospitalizations last anywhere from 3 days to 3 weeks. During these hospitalizations, powerful antibiotics are given intravenously and airway clearance is performed more frequently to “clean out” the infected and weakened lungs.

During clinic visits, a pulmonary function test (PFT) is performed to assess the health of the lungs. During a PFT, patients are asked to blow as powerfully and as long as they can into a machine that measures things like FEV (forced expiratory volume).

