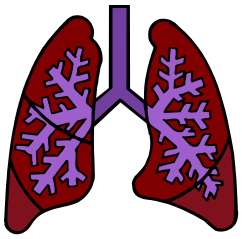
A Disease Process Module:
Understanding Cystic Fibrosis



IN THE KNOW

The Inservice Club for Nursing Assistants

We hope you enjoy this Inservice, prepared especially for nursing assistants like you. You work very hard, and we appreciate the effort you make to complete these educational materials. It shows your desire to continue learning and growing in your profession!

After finishing this inservice, you'll be able to:

- Name the parts of the body that can be affected by cystic fibrosis.
- Name and describe at least three common symptoms of CF.
- Discuss at least 3 possible complications of cystic fibrosis.
- Describe at least 6 ways to help your CF clients stay safe and healthy.

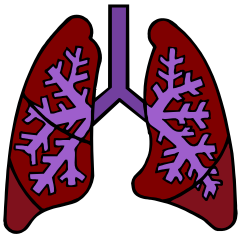
Instructions for the Learner

If you are studying the inservice on your own, please:

- Read through all the attached materials. You may find it useful to have a highlighting marker nearby as you read. Highlight any information that is new to you or that you feel is especially important.
- If you have questions about anything you read, please ask _____.
- Take the quiz. Think about each statement and circle the best answer.
- Check with your supervisor for the right answers. You pass the quiz with at least eight correct answers! Print your name, write in the date, and then sign your name.
- Keep the inservice information for yourself, and turn in the quiz page to _____ no later than _____.
- Show your Inservice Club Membership Card to _____ so that it can be initialed.

THANK YOU!

A Disease Process Module:
Understanding
Cystic Fibrosis



IN THE KNOW

The Inservice Club for Nursing Assistants

Fast Facts About Cystic Fibrosis

- CF is <u>not</u> contagious! *(A person can't catch it like a cold or the flu.)*
- CF is <u>inherited,</u> which means that it's passed down from parents to their children.
- CF is one of the most common inherited diseases.
- The disease is most common in Caucasians.
- CF is usually diagnosed before age 3.
- "Cystic fibrosis" means "fluid-filled sacs with scar tissue."
- CF is a ***fatal*** disease.

"Woe to that child which when kissed on the forehead tastes salty. He is bewitched and soon must die."

This 200-year-old saying comes from Northern European folklore. Although people back then didn't realize it, they were talking about a child with cystic fibrosis. How do we know? Because salty skin is a symptom of the disease.

What is Cystic Fibrosis?

- Cystic fibrosis *(or CF for short)* is a <u>*progressive*</u> and <u>*chronic*</u> disease of the body's mucus glands. (Mucus glands produce tears, sweat, saliva and digestive juices.)
- Cystic fibrosis is <u>*progressive*</u> because it gets worse over time and is *not* reversible.
- CF is <u>*chronic*</u> because it continues over a lifetime.
- Cystic fibrosis affects the mucus of the respiratory system and the digestive system.
- Normal mucus is *thin* and *slippery*. But with CF, the mucus is *thick*, *sticky*—and *deadly*. It plugs up tubes, ducts and passages, especially in the lungs, intestines and pancreas.
- For many, many years, cystic fibrosis killed children while they were still infants or toddlers. By 1960, doctors were able to help most CF kids live until age 7.
- In the 1970's, because of improved treatments, many children lived to be young teenagers. And, by the late 1980's, kids with CF could expect to live until age 18.
- In recent years, new ways have been discovered to help people with CF. Now, the victims of CF are living longer, fuller lives!

Keep reading to learn more about cystic fibrosis.

How Much Do You Know About the Lungs?

Just take a guess...then turn this page upside down to see the answers!

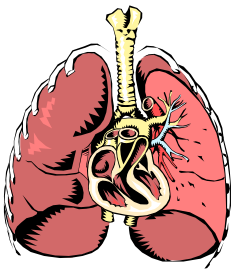
1. Together, the average adult lungs weigh about _____?
   - A. *4 lbs.*
   - B. *2.5 lbs.*
   - C. *1 lb.*
2. If the lungs were spread out flat, they would nearly cover a _____.
   - A. *Tennis court.*
   - B. *Tennis racket.*
   - C. *Bowling alley.*
3. The lungs are a _____ color.
   - A. *Green.*
   - B. *Blue/gray.*
   - C. *Pink/gray.*
4. The lungs look sort of like balloons. They are large and _____.
   - A. *Round.*
   - B. *Oval.*
   - C. *Cone-shaped.*
5. The lungs are not shaped exactly the same. The left lung is slightly smaller than the right lung because it has to share some chest space with the _____.
   - A. *Hand.*
   - B. *Heart.*
   - C. *Colon.*

Answers: 1. 2.5 lbs. 2. Tennis court 3. Pink/gray 4. Cone-shaped 5. heart

Interesting Facts About Cystic Fibrosis

- There are about 30,000 people in the U. S. with CF.
- Every year in the U.S., nearly 2,500 babies are born with CF.
- CF is *more* common in whites and *less* common in African Americans, Native Americans and Asian Americans.
- Today, about half of the people with CF live to be thirty years old. (Unfortunately, the other half die at a younger age.)
- In 1938, Dr. Dorothy Anderson, of Columbia University noticed scar tissue on the pancreas during an autopsy. She wrote the first detailed account of how this disease affects the body—and she gave it a name: "cystic fibrosis of the pancreas."

Facts from the National Heart, Lung, and Blood Institute and The Cystic Fibrosis Foundation



The thick mucus caused by CF helps bacteria grow, so people with CF tend to have frequent infections. Over time, these infections damage the lungs, causing respiratory failure. This is the leading cause of death for people with cystic fibrosis.

According to the Cystic Fibrosis Foundation, over 10 million people carry the defective gene that causes CF—and they <u>don't</u> even know it.

Since 1988, the number of <u>adults</u> with cystic fibrosis has grown by 78%! (Why? Because people with CF are living longer!)

What Causes Cystic Fibrosis?

- Cystic fibrosis is known as a genetic disease because it's caused by a defective gene that's passed down from parents to their children.
- So far, scientists know that this bad gene causes CF and that some children are born with this gene. However, they don't know why the gene is defective. They continue to do research to find the answers. *(Check out page 4 for how cystic fibrosis is inherited.)*

In 1989, scientists identified the "bad" gene that causes CF. Its discovery has helped with CF treatments.

How is Cystic Fibrosis Diagnosed?

- Most of the time, CF is diagnosed in children before the age of two. A small number of children are not diagnosed until later on in childhood.
- The most common test for CF is the **sweat test**. This is a painless test that measures the amount of salt in a person's sweat.
- Unfortunately, this test can't tell if the cystic fibrosis is *mild* or *severe*.
- Because newborns produce very little sweat, the sweat test isn't useful for them. As a result, doctors don't usually perform the test on infants younger than four months old.
- Here are more tests that help doctors diagnose CF:
  - Blood tests.
  - Chemical genetic tests.
  - Chest x-rays.
  - Lung function tests.
  - Phlegm (mucus) cultures.
  - Stool tests.

The sweat test was developed over 40 years ago by a scientist who noticed that CF patients have really salty sweat. It's still used today.

A Few Myths About Cystic Fibrosis

MYTH: You can catch CF like a common cold.

FALSE: Since clients with CF have a chronic cough, it makes other people think germs are being spread. However, CF is an *inherited* disease—it's not contagious.

MYTH: CF causes brain damage and mental retardation.

FALSE: Sometimes people get *CF* mixed up with *CP* (cerebral palsy). CF does not affect the brain or cause intellectual disabilities.

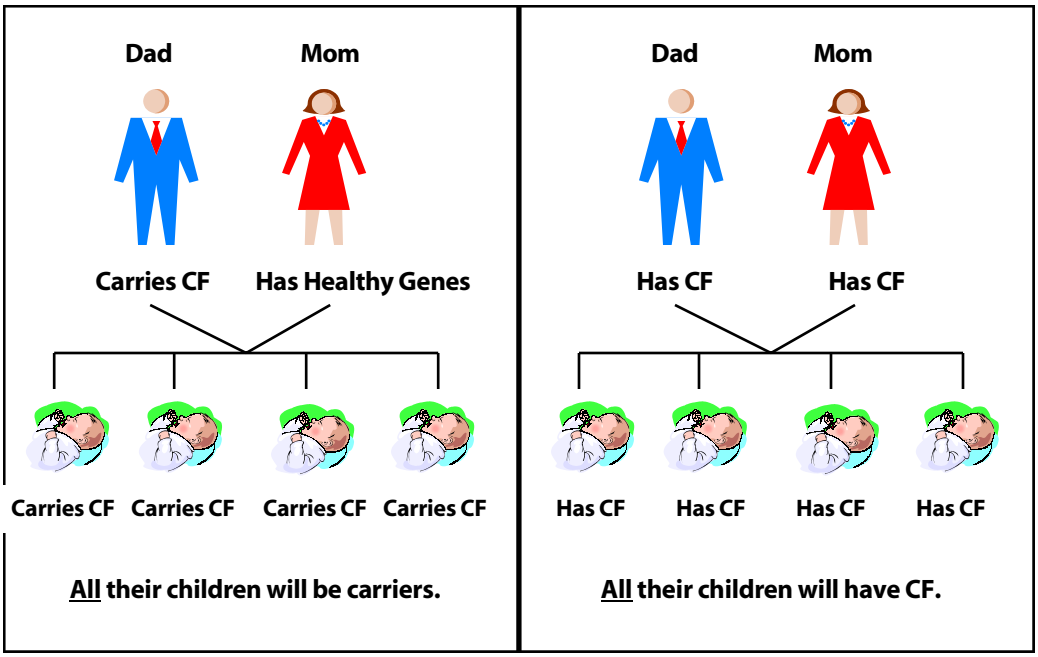
MYTH: CF is caused by something the parents did during pregnancy.

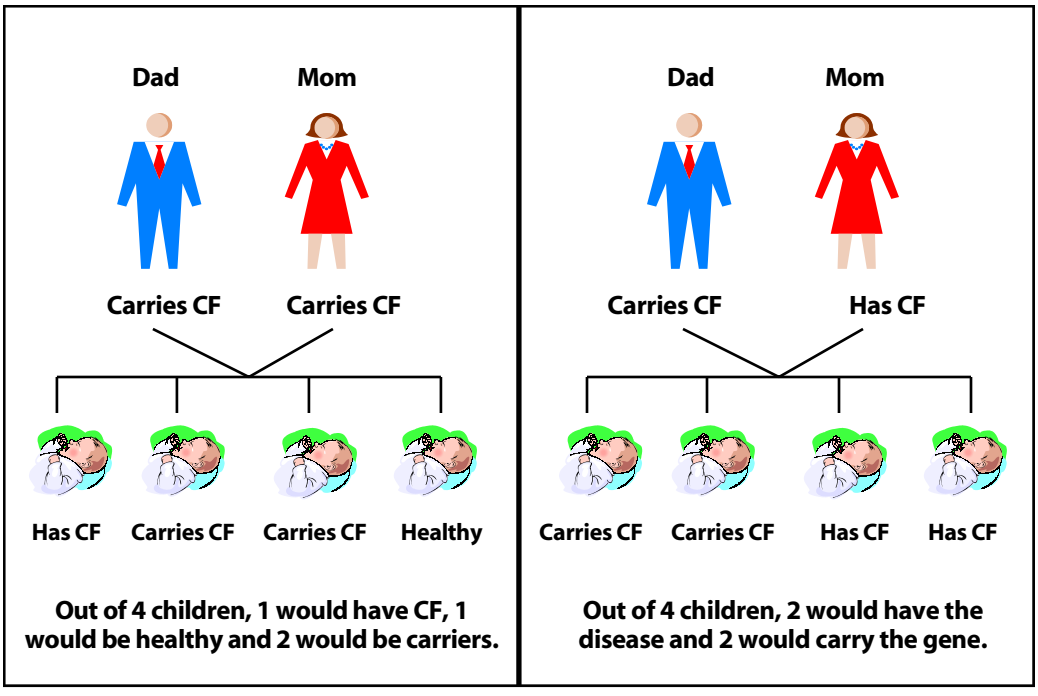
FALSE: Some parents may feel guilty when their children are born with CF, but it isn't their fault. CF is caused by a faulty gene over which they have no control.

Why do people with CF cough so much? Thick and sticky mucus gets stuck in their air passages making it very hard to breathe. Coughing loosens up the mucus.

The Cystic Fibrosis Heredity Factor

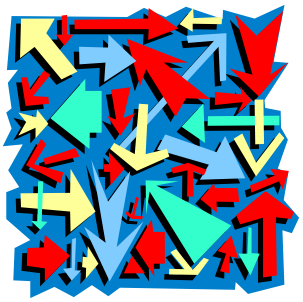
- To develop CF, a child *must* inherit <u>two</u> defective genes—one from each parent.
- People with CF have the defective gene. So do "carriers"—although they never get sick themselves.
- The four possible inheritance patterns are shown below:





Pregnancy is risky for women with CF. However, if a woman has good lung function, she has a chance for a healthy baby.

Cystic Fibrosis can happen to both boys and girls because the CF gene is <u>not</u> linked to the gene that decides a baby's sex.



A parent who carries the CF gene may have a child with *mild* or *severe* CF. The same is true for a parent who has the disease.

Most Common Symptoms of Cystic Fibrosis

- CF can affect many different organs in the body including the lungs, intestines, sweat glands, pancreas, sinuses, liver, gallbladder and the reproductive system. Signs and symptoms depend on which part of the body is affected.
- Not all your CF clients will have the same symptoms. Some clients may have severe "full-blown" CF while others may have milder symptoms. For example, one child may have lung problems, but not digestive troubles. Another child may have both. Symptoms vary as children get older, too.

1. Too Much Salt

- People with CF make excessive amounts of salt. They tend to have 2 to 5 times more salt in their sweat than people without CF.
- Excess salt causes the skin to taste salty. Some people say that CF causes a salty "frosting" on the skin.
- People with cystic fibrosis lose a lot of salt when they sweat. This can be a big problem for them. It can cause abnormal heart rhythms and puts them at risk for going into shock.

2. Thick Mucus

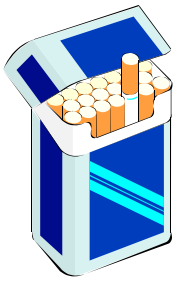
- CF causes people to have very thick, sticky mucus that builds up and clogs passages in the body—*especially in the lungs and intestines*.
- In the lungs, the mucus plugs up the airways, making it hard to breathe. The extra mucus gives bacteria a great place to grow.
- In the pancreas, thick mucus stops digestive juices from getting to the intestines. Thick mucus can also cause dangerous blockages in the intestines.

3. Chronic Cough

- Most people with CF have a chronic cough caused by thick mucus and frequent respiratory infections.
- When people with CF cough, they may produce a heavy, discolored mucus— *or even blood*.
- Sometimes the coughing becomes very violent , which can be exhausting and painful.
- Wheezing can also be a problem, especially for someone who has both asthma and CF.

People with cystic fibrosis have mucus so thick that it's been described as "*thicker than a milkshake.*"

Bad breath can be a problem for people with cystic fibrosis due to their frequent respiratory infections and poor digestion.



A father once described his young son's cough in this way—*"It sounds like he has a pack-a-day smoking habit."*

Most Common Symptoms of CF *(continued)*

4. Respiratory Infections

- Since thick mucus is a breeding ground for bacteria, people with CF tend to get a lot of respiratory infections in their lungs and sinuses. (*See pages 7 and 11 for more on this*.)
- These constant infections lead to symptoms of poor lung function, including: shortness of breath, difficulty breathing and a bluish color to the skin and/or fingernails. These symptoms drain CF clients of their energy.
- Gradually, these infections cause damage to the lungs. The damage may be so bad that a lung transplant is needed.

When CF symptoms get worse, children are often admitted to the hospital. By age 10, some children have been admitted 30 times or more!

5. Foul Smelling Stools

- Most people with CF don't produce enough digestive juices. This causes them to have foul smelling, greasy, bulky stools.
- When people with CF eat fat, it passes through their bodies undigested. This causes their bowel movements to be filled with fat.
- They tend to have *frequent* bowel movements and chronic diarrhea.
- They may also suffer from a swollen belly, abdominal pain and gas.

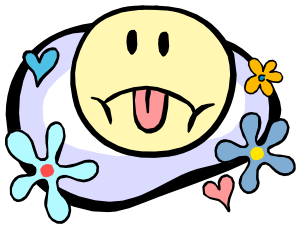
6. Poor Growth

- Because people with CF have problems with digesting food, they tend to have trouble keeping weight on.
- Their bodies are unable to absorb either fat or basic vitamins. This keeps them from gaining weight and growing properly—even if they have good appetites.
- If they aren't treated for this problem, people with CF develop symptoms of <u>starvation</u>.

7. Reproductive Problems

- Cystic fibrosis affects the reproductive organs of both sexes. Sexual development is delayed for about two years.
- 95% of males with CF aren't able to have children because certain parts of the reproductive tract are absent or underdeveloped.
- Females with CF may have a harder time getting pregnant because of abnormal mucus and menstrual irregularity.

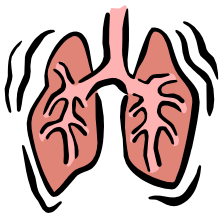
Dr. Fielder of Cincinnati Children's Hospital says, "Infants with CF are hungry, irritable, and colicky all the time."



For healthy people, about 3 gallons of digested food go through the digestive system every day. People with CF have a hard time with this process.

Watch Out For These Complications of CF ...

- **Pneumonia** is an inflammation and infection of the lungs. There are more than 50 kinds—some mild and some serious. Pneumonia causes the lungs to become infected making it hard to breathe.
- **Bronchitis** is an infection of the bronchial tubes in the chest. It often follows another illness such as a cold.
- **Polyps** are round, fleshy growths inside the nose.
- **Clubbing** is an enlargement, rounding and /or swelling of the fingertips and toes caused by lack of oxygen.
- **Sinusitis** is an inflammation and infection of the cavities behind the nose. It causes congestion in the nasal passages.
- **Collapsed Lungs** occur when a hole in the lung tissue traps air between the lung and the chest wall. This can become a chronic problem.
- **Hemoptysis** is the coughing up of blood. Bright red blood means *new* or *active* bleeding. Off-red or brown blood means *old* bleeding.
- **Rectal Prolapse** happens when part of the rectum pushes through the anus. It's often caused by intense coughing or by problems having bowel movements.
- **Liver Disease** is an inflammation of the bile ducts. It can cause serious health problems.

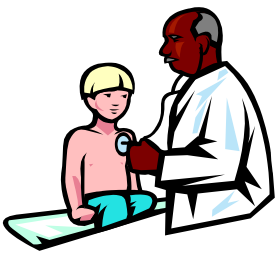


Each year, more than 40,000 Americans die of pneumonia. So, it's important to get treatment right away.

In some newborns, the first symptom of CF may be an intestinal blockage called "meconium ileus". This happens when the tarry greenish-black stool (meconium) that's usually passed by the baby within a few days of birth becomes so thick that it gets stuck.

More Common Complications of CF ...

- **Asthma** often develops along with CF—usually due to allergies.
- **Diabetes** can develop as CF destroys the pancreas.
- **Osteoporosis** develops due to a lack of calcium.
- **Cor pulmonale** is an enlargement of the right side of the heart.
- **Bowel blockages** can develop when stool is unable to pass through the intestine because of thick, sticky mucus.
- **Gallstones** are small round "pebbles" in the gallbladder. They are usually made of calcium.
- **CF Arthritis** causes joint aches and pains that seem to happen when the lungs are in bad shape.



About 10% of children with CF become diabetic after age 8 because of damage to the pancreas.

Can Cystic Fibrosis Be Treated?

So far, there is no cure for CF. But there are treatments that help reduce symptoms, slow down the progress of the disease and make living with CF easier. These treatments include:

- **Antibiotics.** These medications help control respiratory infections.
- **Mucolytics.** These are mucus-thinning drugs which help clear up lung and intestinal blockages.
- **Decongestants.** These drugs reduce swelling of the breathing tubes.
- **Bronchodilators.** These medications are mist sprays that come in inhalers. They help with breathing by widening the bronchial tubes.
- **Enema.** An enema may be necessary to help treat an intestinal blockage.
- **Vitamins/Diet.** People with CF need 100% more calories than healthy people. They need a diet that is:
  - Well-balanced
  - High-calorie
  - Low-fat
  - High-protein
- **Pancreatic Enzymes.** These pills help digest food.
- **Chest PT.** People with CF need the mucus drained from their lungs at least twice a day. This can be done by clapping them on the front and back of the chest or by using a special device like the "flutter." (The "flutter" is a small, hand-held device that looks like a pipe. You blow into it.)
- **Lung Transplant.** This treatment is usually the last resort.

Alternative therapies, like acupuncture or herbal remedies, are becoming popular to help people with CF control their symptoms. However, remind your clients to check with their doctors first!

There are 4 main goals for treating cystic fibrosis:

1. *Treat infections.*
2. *Keep lungs free of mucus.*
3. *Help air flow through the lungs.*
4. *Provide enough calories and nutrition.*

Famous People Who Have Relatives with CF

- Celine Dion—*singer, had a niece who died from CF in 1993 at age 16.*
- Rosie O'Donnell—*talk show host, has a nephew with CF.*
- Boomer Esiason—*football quarterback, has a son with CF.*
- Kimberly Myers—*NASCAR racer, has CF.*
- Alfred Hitchcock—*film director, had a granddaughter with CF who had a lung transplant.*
- Jimmy "The Greek" Snyder—*mafia member, had sons with CF.*

Right now in the United States, CF is the number one killer of children who have an inherited disease.

Some Tips for Helping Clients with CF

- Keep an eye on your clients as they take their medications, *especially inhalers*. It's important that they know how to use them properly. Let your supervisor know right away if you think a client may be misusing an inhaler.

- Using an inhaler *seems* simple, but you would be very surprised at how many people forget to take off the cap! Most people don't use an inhaler correctly and when it's used the wrong way, less medicine gets to the lungs. To use an inhaler, CF clients should:
  - Shake inhaler for five seconds and remove the cap. Breathe out all the way. Hold inhaler one or two inches in front of mouth. Breathe in slowly through mouth and press down on inhaler one time. Hold breath and count to ten.
  - If using a spacer or holding chamber, first press down on inhaler, then wait five seconds *before* breathing in. *(Spacers allow the medicine to get deep into the lungs. They are usually recommended for children and people who have trouble using their inhalers correctly.)*

- Ask to see your client's inhaler. If you see a "powder" around the hole where the medicine comes out, the inhaler needs to be cleaned. You do this by removing the medication canister from the mouthpiece and rinsing the mouthpiece and cap in warm water. It's best to do this in the evening so the mouthpiece can "air dry" overnight.

- Not only is it important to watch how your clients are using their inhalers, it's a good idea to watch how they use chest PT devices like the "flutter."

- Remind your clients to always take their medications as prescribed by their doctor—no more or no less! (This includes oxygen and vitamins, too.)

- Be alert! Let your supervisor know right away if your CF clients are mixing their medications with other prescription drugs, and/or over-the-counter drugs. Mixing medications can cause serious, unexpected side effects.

- Also, remind your clients not to share any of their medications with other people.

- If your CF client is a young child and dislikes taking medications, share these hints with the child's parent or nurse: *Roll the pill in some soft butter just before the child swallows it. If a pill gets stuck, offer a bit of banana to help push it down. Mix a pill in some yogurt or pudding, then put it on a spoon for the child to swallow.*

- Suggest that your CF client set up a routine for therapy and always stick to it. It's helpful to do the sessions at the same time each day.

Remember…

*If you have any questions about what **you can** or **can't do** for a client with cystic fibrosis, ask your supervisor.*

Keep in mind that many CF clients will be young children. They may not be as willing to take their medicines or do their chest physical therapy. Tell your supervisor if they are skipping their meds and/or therapy sessions.

Watch your clients for worsening of symptoms. Call your supervisor right away if you think symptoms have changed in any way!

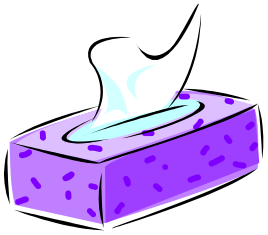
More Tips For Helping Clients with CF

- Some of your clients may use oxygen. If so, make sure the equipment is used correctly. Call your supervisor if your client seems to need help.
- Help your clients change their oxygen nose tubes often— especially if the prongs become dirty or uncomfortable.
- If your clients have portable oxygen units, make sure they know exactly how much oxygen they have left. You don't want them to run out unexpectedly.
- It's important not to smoke around oxygen, and it's *very* important not to smoke around clients with CF. Encourage your clients not to smoke at all.
- Suggest that your CF client get an annual flu shot and the pneumonia vaccine.
- Infection control is important, too. People with cystic fibrosis are more at risk than healthy people for catching—*and getting seriously ill from*—sicknesses such as colds and flu.
- Remember to wash your hands often. In addition, encourage your client's family members to wash their hands before visiting to get rid of any germs they may have "picked up" during the day.
- Remind and/or help your CF clients to wash their hands frequently, too, especially if they are sick with a bacterial infection.
- Suggest that your clients wear masks over their noses and mouths when visiting the doctor's office, hospital or anyplace where there may be a lot of germs.
- Encourage your CF clients to lead as normal and active a life as possible.
- Regular exercise helps loosen the mucus in the airways, stimulates coughing to clear the mucus and strengthens the lungs and heart. For many people with CF, participating in an exercise program or sport helps keep the body in good health while improving confidence and self-esteem.
- Always make sure that exercise is *ordered* for a client before you suggest it—even if it's just walking around the block.
- Exercise should be fun! Your client may enjoy it more if you do it together. Take a walk around your client's home, neighborhood, or within your facility. It's more fun with a "buddy."

Second-hand smoke is harmful for everyone—especially people with CF because their lungs are already stressed. Cigarette smoke makes it even harder for them to get rid of mucus.

Notify your supervisor if your CF client has :

- *A fever.*
- *Breathing difficulties.*
- *Increased tiredness.*
- *Lack of appetite.*



Some people with CF suggest putting pieces of tissue or cotton balls between oxygen nose tubes and the skin to prevent rubbing.

Even More Tips for Helping Clients with CF

- Depression is common among people with a chronic disease. Watch for: *weight loss or gain, lack of energy, anxiousness, overwhelming feelings of sadness, sleeping more than usual, loss of interest in activities* and *thoughts of suicide.*
- Encourage your clients to drink plenty of fluids (about 8 glasses a day) to avoid dehydration and to loosen up mucus.
- Good nutrition is crucial! Check each client's care plan for special dietary needs and encourage all clients to eat well-balanced meals.
- Because their bodies can't digest properly, people with CF have to take pancreatic enzymes and a number of vitamins. Remind your clients to take their enzyme medications and vitamins as ordered.
- Remember that your clients may have problems digesting fats. However, if your clients are able to digest fat, encourage high fat foods like cheese, cream, butter, whole milk and peanut butter. (These are all high-calorie foods, too!)
- Because people with cystic fibrosis *lose* a lot of salt, encourage your clients to eat salty foods and to add as much salt as possible to their meals.
- If you cook for your clients, try to cook in ways that increase calories, including:
  - *Sauté potatoes and vegetables.*
  - *Offer fried rice.*
  - *Cook pasta with oil in the water.*
  - *Fry meats instead of grilling them.*
- To help your clients increase their intake of protein and calories, suggest that they:
  - *Eat whenever they are hungry.*
  - *Keep snack foods handy—such as cheese and crackers, muffins, trail mix and pretzels.*
  - *Eat on a regular basis—don't skip meals!*

Most CF clients have repeated respiratory infections like pneumonia and bronchitis. Watch for signs of both.

Symptoms of Pneumonia	*Symptoms of Bronchitis*
• *Shaking chills*	• *Soreness in chest*
• *High fever*	• *Breathlessness*
• *Sweating*	• *Wheezing*
• *Chest pain*	• *Slight fever*
• *Cough—with thick, rust-colored, greenish or yellow phlegm*	• *Cough that may or may not have yellowish-gray or green sputum*

Encourage your clients to see their doctors, therapists, dietitians and social workers on a regular basis. It takes a "team effort" to help people with CF live full and longer lives. Don't forget that you are part of that team!

High-calorie, high-protein, and low-fat drinks like Lipisorb, Equate, Nutra-Shake, and Equate Plus can help a person with CF maintain their weight.

It's recommended that CF clients avoid drinking alcohol. Alcohol slows down the respiratory system and adds stress to the liver.

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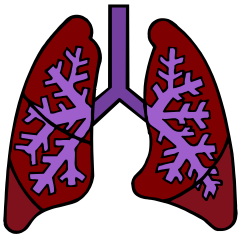

EMPLOYEE NAME *(Please print)*:

_____

DATE: _____

- ***I understand the information presented in this inservice.***
- ***I have completed this inservice and answered at least eight of the test questions correctly.***

EMPLOYEE SIGNATURE:

_____

SUPERVISOR SIGNATURE:

_____

Inservice Credit:	
☐ Self Study	1 hour
☐ Group Study	1 hour

File completed test in employee's personnel file.

IN THE KNOW

The Inservice Club for Nursing Assistants

Are You "In the Know" about Cystic Fibrosis?

Circle the best choice and then check your answers with your supervisor!

1. Your CF client, Danny, doesn't like using his inhaler. You notice he skips doses. You should:

A. Let his mother handle the situation.
B. Clean his inhaler for him.
C. Tell your supervisor that he is skipping doses.
D. Offer to give him candy if he stops skipping doses.

2. True or False
Cystic Fibrosis is a contagious and curable disease.

3. A baby with CF may need more diaper changes than normal because:

A. The baby drinks too many fluids.
B. The baby is always crying.
C. The baby's frequent stools smell terrible, and may irritate the skin.
D. The baby eats too much.

4. True or False
Cystic Fibrosis affects everyone in the same way.

5. Lizzy complains that she constantly coughs when she rides her bike. You should:

A. Suggest that she take some cough syrup.
B. Tell her to stop riding her bike.
C. Remind her that exercise and coughing are good ways to bring up the mucus.
D. Encourage her to watch T. V. instead.

6. People who have cystic fibrosis may have trouble with:

A. Respiratory infections.
B. Chronic coughing.
C. Poor growth.
D. All of the above.

7. True or False
The leading cause of death in CF clients is heart failure.

8. True or False
Cystic fibrosis is inherited—meaning that it's passed down through families.

9. True or False
Cystic fibrosis is a disease of the bones.

10. True or False
Most people who have cystic fibrosis die before age 10.