UNDERSTANDING CYSTIC FIBROSIS

INTRODUCTION

Cystic fibrosis is a chronic disease that affects the respiratory system, the pancreas, the gastrointestinal system, and many other organs. People with cystic fibrosis have a genetic defect that causes production of an excess amount of thick viscous mucous, and this results in damage to specific organ systems and serious respiratory, metabolic, and gastrointestinal complications.

There is no cure for cystic fibrosis, but medications, physical therapy, diet, and lifestyle adjustments can help prevent complications and make this disease easier to live with. However, cystic fibrosis is a very serious disease. Patients often require a lot of care and support, and the prognosis for someone with cystic fibrosis is not good. The average life expectancy of people who have cystic fibrosis is much, much lower than that of the average American.

Learning Break: The name cystic fibrosis was given to this disease many years ago because people with cystic fibrosis have cysts (small closed sacs) in the pancreas and fibrosis in the pancreas and the lungs.

STATEMENT OF PURPOSE

This module is intended to provide certified nursing assistants (CNAs) with information about: the pathophysiology of cystic fibrosis; the signs and symptoms of the disease; treatment options, and; the basic process of care for someone who has the disease.

WHAT CAUSES CYSTIC FIBROSIS?
Cystic fibrosis is a genetic disease; the tendency to develop cystic fibrosis is inherited. Some people who have the gene that can cause cystic fibrosis only have one copy of this gene. However, if someone inherits a copy of the defective gene from each parent then that person will develop cystic fibrosis.

The gene involved in cystic fibrosis is responsible for the production of mucous in the lungs, the pancreas, the gastrointestinal tract, and other areas. Mucous is a naturally occurring compound that helps keep the lungs warm and moist and acts as a lubricant and as a protective barrier in the stomach, intestines, and other organs. Mucous also traps bacteria and viruses and prevents them from causing infections and in parts of the gastrointestinal tract mucous helps digest food and helps move digested food through the gut.

For people without cystic fibrosis mucous is relatively thin and watery. But in cystic fibrosis, the gene that regulates mucous production is abnormal and people who have this disease produce mucous that is unusually thick, viscous, and sticky; it tends to adhere to the walls of the lungs and the gut. The abnormal mucous can block the bronchi and other air passages in the lungs, and it can also block the gastrointestinal tract in various places such as pancreas, the gall bladder, and the bowels. This thick, sticky mucous also is an excellent place for bacteria to grow and because of that people who suffer from cystic fibrosis often have chronic infections and inflammation, especially in the lungs and the gastrointestinal tract.

**Learning Break:** Genes are the part of a cell that contain the information that regulates how cells reproduce and determines what their structure and functions
will be. In cystic fibrosis, the genes involved control the cells that produce mucous, but the genetic information is faulty and the mucous that is produced, as mentioned above, is thick and viscous and very sticky. Cystic fibrosis is an autosomal recessive disorder. That means that in order for these abnormal genes to express themselves and to cause the mucous-producing cells to make abnormal mucous, there must be two of these genes present, one from each parent. Genes are found in pairs and for a disease such as cystic fibrosis to occur, both of the genes that control the production of mucous must be abnormal.

**RISK FACTORS: WHO GETS CYSTIC FIBROSIS**

Cystic fibrosis is the most common lethal inherited disease in the white population. In the United States, cystic fibrosis affects approximately 1 of 3500 Caucasians, 1 of 9500 Hispanic Americans, 1 of 17,000 African Americans, and 1 of 31,000 Asian Americans. Males and females are equally affected, but females who have cystic fibrosis have more severe lung problems and die at a younger age. The signs and symptoms of cystic fibrosis usually begin in childhood, often by the age of 6-8 months, but in about 5% of people who have cystic fibrosis the disease developed when they were adults.

**SIGNS, SYMPTOMS AND COMPLICATIONS OF CYSTIC FIBROSIS**

Cystic fibrosis is primarily a disease that affects the lungs and the pulmonary problems of cystic fibrosis will be discussed in detail. However, cystic fibrosis can also have serious adverse effects on many other organ systems.
Respiratory

Cystic fibrosis involves many of the body’s organ systems but the lungs are of first concern. The mucous in the lungs of people who have cystic fibrosis is very thick, viscous, and sticky. There is also an abnormally large amount of mucous produced. This has many severe implications.

In the normal lungs, mucous gets removed in several ways. Coughing and sneezing help expel some mucous and coughing and sneezing both move a large amount of mucous to the trachea and the oral cavity, at which point it can be expelled and/or swallowed. Mucous is also moved out of the respiratory tract by the cilia. The cilia are tiny hair-like fibers located in the walls of the left and right main stem bronchial passages and in the trachea. The cilia move in a wave-like fashion that physically pushes mucous in the respiratory passages up and out towards the trachea and the mouth. At that point the mucous can be removed by involuntary and voluntary coughing, sneezing or as mentioned previously, by swallowing. This process is called the mucous escalator.

However, because the mucous in the lungs of someone with cystic fibrosis is so dense and thick, because it adheres so tightly to the airway passages, and because there is such a large amount of mucous being produced, coughing, sneezing, and the mucous escalator cannot effectively remove it and the excess mucous stays trapped in the lungs.

**Learning Break:** The terms mucous, phlegm, and sputum are often used interchangeably when discussing cystic fibrosis. Phlegm is abnormally thick.
mucous produced in the respiratory tract, and sputum is mucous in the respiratory tract

   The small and medium-sized airway passages that are deep in the lungs are the most affected and the large amount of concentrated mucous that is produced harms the lungs in several ways.

   1. The mucous forms a barrier, plugging the small and medium-sized airways and preventing oxygen from moving from the lungs into the bloodstream.

   2. The mucous causes irritation and inflammation of the small and medium-sized airways.

   3. Inflammation eventually causes damage to these airways.

   4. The mucous provides an ideal environment for bacteria to grow and bacterial lung infections are very common in people who have cystic fibrosis. The great majority of people who die from cystic fibrosis die from a lung infection.

   The typical respiratory signs and symptoms of cystic fibrosis are **cough**, **difficulty breathing** at rest or during physical activity, and **wheezing**. The excessive mucous production prevents oxygen from moving through the lungs, so people who have cystic fibrosis often feel short of breath. The excessive and excessively thick mucous is very irritating to the respiratory passages and in response, people with cystic fibrosis develop a persistent cough to try and clear the secretions. Cystic fibrosis can also cause chronic sinus congestion and sinus infections.
Diabetes Mellitus and Cystic Fibrosis

Diabetes mellitus is a common complication of cystic fibrosis. There are three ways that cystic fibrosis causes diabetes.

1. Pancreatic damage: The pancreas produces digestive enzymes that are used to physically break down food in the gut. These enzymes are transferred from the pancreas to the gastrointestinal tract through the pancreatic duct. Excess mucous production blocks the release of pancreatic digestive enzymes through the pancreatic duct and these enzymes, instead of digesting food, begin to digest the pancreas. Eventually the islet cells that produce insulin are irreversibly damaged and there is no insulin available.

2. Insulin release: When the islet cells of the pancreas are functioning normally an elevated blood sugar triggers the release of insulin so that blood sugar can be returned to a normal level. Cystic fibrosis prevents the release of insulin when it is needed. This is a relatively new discovery about cystic fibrosis and diabetes mellitus.

3. Insulin resistance. Insulin resistance is a condition in which the body does not respond to insulin. The insulin levels are normal (or more often, abnormally high) but blood sugar is not lowered as would be expected. Insulin resistance is the common cause of type 2 diabetes.

In these ways the diabetes caused by cystic fibrosis is similar to type 1 diabetes in that no insulin is produced, and it is similar to type 2 diabetes in that insulin is produced but it does not function properly to lower blood sugar.
Diabetes mellitus caused by cystic fibrosis usually begins to develop in the late teens and early 20s and more than 50% of adults who have cystic fibrosis have diabetes mellitus. Women are affected more often than men. The presence of diabetes mellitus in people who have cystic fibrosis is associated with an increased incidence of lung infections, a decreased rate of survival, and an increased progression of cystic fibrosis itself. Oral medications are not effective for these patients; they must be treated with insulin.

Gastrointestinal Disease and Cystic Fibrosis

People who have cystic fibrosis typically suffer from abdominal cramps, constipation, obstructions, nutritional deficiencies and weight loss. These signs and symptoms happen for two reasons: 1) the excess, thick mucous in the digestive tract can block the bowels, it can block the bile ducts of the gallbladder, and it can block the drainage ducts of the pancreas, and; 2) the mucous provides an environment that allows bacteria to grow and the gastrointestinal tract and its organs can become inflamed. Because of these pathologic processes, people who have cystic fibrosis are prone to various diseases of the gastrointestinal tract and complications related to malabsorption. Some of these are briefly discussed below.

1. Gallbladder and nutritional deficiencies: Bile is a compound that is produced in the liver and stored in the gall bladder. Bile is secreted from the gall bladder into the bowels through a passage called the bile duct and bile is necessary for the absorption and digestion of fats and fat-soluble vitamins. As with the pancreas, excess amounts of thick,
adherent mucous block the bile duct and prevent bile from reaching the intestine. Because of this process people with cystic fibrosis can suffer from fat malabsorption and vitamin deficiencies of the fat-soluble vitamins, vitamins A, D, E, and K. People who have cystic fibrosis are also susceptible to gall stones and gall bladder infection.

2. Bowel obstruction: Infants who have cystic fibrosis are likely to develop a specific type of bowel obstruction called meconium ileus. Meconium is the stool that is in a baby’s gastrointestinal tract when she/he is born and meconium is very thick and tenacious. Bowel obstructions can also occur in other parts of the gastrointestinal tract in adults.

3. Bone loss: Calcium is a mineral that is considered to be the “building block” of the bones: calcium is what gives bones their density and hardness. We depend on dietary sources for our calcium but calcium cannot be absorbed without vitamin D. Because vitamin D absorption is negatively affected by cystic fibrosis, people who have the disease are susceptible to osteoporosis and bone fractures. Approximately 75% of all people who have cystic fibrosis have evidence of osteoporosis. Bone loss in these patients can also be caused by medications used to treat the pulmonary complications of cystic fibrosis and by lack of exercise.

4. Cirrhosis of the liver: Cirrhosis is a medical term that means that normal tissue has been replaced by fibrous scar tissue. Cirrhosis of the liver can be caused by cystic fibrosis when the liver ducts that transport
bile to the gall bladder become obstructed with mucous and become inflamed.

Reproduction and Cystic Fibrosis

Approximately 95% of men who have cystic fibrosis are infertile. Infertility is less common in women who have cystic fibrosis but it still affects approximately 50% of this population. Infertility in men who have cystic fibrosis is caused by structural abnormalities of the reproductive tract; in women it is complex and multi-factorial.

Other Complications of Cystic Fibrosis

Other complications of cystic fibrosis include fluid and electrolyte imbalances; heart failure; kidney stones; reflux disease, and; rickets.

Table 1: Signs and Symptoms of Cystic Fibrosis

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<tr>
<th>Abdominal pain</th>
<th>Constipation</th>
<th>Cough</th>
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<td>Decrease exercise tolerance</td>
<td>Diarrhea</td>
<td>Dry skin</td>
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<td>Dyspnea</td>
<td>Fatigue</td>
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<td>Hyperglycemia</td>
<td>Tachypnea</td>
<td>Weight loss</td>
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<td>Wheezing</td>
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HOW IS CYSTIC FIBROSIS DIAGNOSED?

Cystic fibrosis is diagnosed using the following three methods:

- Genetic testing: Genetic testing to detect cystic fibrosis can be done in the pre-natal period or post-natally. The pre-natal genetic screening tests are
very sensitive and very accurate. Post-natal newborn screening for cystic fibrosis is required by law to be done on all babies. A blood sample is obtained soon after birth and examined for the genetic abnormality that causes cystic fibrosis.

- **Sweat test:** People with cystic fibrosis produce sweat that has a very high sodium (salt) content. If the initial test shows high sodium content, the test is repeated several weeks later for confirmation. If the sweat test is negative but the patient has some signs of cystic fibrosis the test should be repeated or if the sweat test result is considered to be borderline the test should be repeated.

- Examining the patient for the presence of respiratory and pancreatic complications that are typical of cystic fibrosis.

**PROGNOSIS FOR CYSTIC FIBROSIS**

There is no cure for cystic fibrosis and unfortunately a diagnosis of cystic fibrosis is accompanied by a very disheartening prognosis. Not all cases of cystic fibrosis are severe. There are variations in the presentation and some people who have cystic fibrosis have a mild form of the disease. But cystic fibrosis is for the great majority of people a very serious disease. Advances in treatments have improved the outlook - more than half the people with cystic fibrosis are 18 years or older - but 20% of the people who have cystic fibrosis do not live to adulthood. Cystic fibrosis is usually diagnosed by age 6-8 months and even with advancements in treatments, the median life expectancy is approximately 40 years.
Learning Break: Median is a statistical term that indicates that 50% of a specific group, population or numerical result is less than the median point and 50% is greater. In this case it indicates that approximately one-half of all patients who have cystic fibrosis will die before age 40 and one-half will survive past that age, for how long it is not known.

Diagnosis at a very young age, severe symptoms, rapid progression of the disease, the presence of diabetes mellitus, and the complexity of a case are factors that contribute to a poor prognosis. Some patients who have cystic fibrosis may survive into their 50s and this is a far better outlook than what was expected not so long ago. In the 1950s, for example, many patients with cystic fibrosis died in childhood. However, no one who has cystic fibrosis will live to the average life expectancy of most Americans.

Treatment of cystic fibrosis involves diet, exercise, medications, physical therapy and occasionally, surgery. The treatment of this disease truly requires a team approach and patients are best served by professionals who specialize in the disease. There are cystic fibrosis centers in the United States that provide state of the art care; a list of these can be found on the website of the Cystic Fibrosis Center at [www.cff.org](http://www.cff.org). The treatment of cystic fibrosis also requires intense involvement of the patient and her/his family.

TREATMENT OF CYSTIC FIBROSIS

There is no cure for cystic fibrosis and living with this disease is very difficult. The three main goals of treating cystic fibrosis are 1) maintaining healthy lung function; 2) maintaining good nutrition, and 3) managing complications. All of
these are important but maintaining healthy lung function is perhaps the most important because respiratory infections are the most common cause of death in patients who have cystic fibrosis.

**Diet and Cystic Fibrosis**

A nutritional consult and period follow-up with a nutritionist is very important for patients who have cystic fibrosis. These patients should be encouraged to eat a high-energy diet that has extra fat, extra fat-soluble vitamins, and extra calories. Patients should be routinely monitored for fat-soluble vitamin deficiencies. Aside from those recommendations, the patient who has cystic fibrosis does not need a highly specialized or restrictive diet unless he/she has diabetes. Good hydration is also important because dehydration can increase the thickness of mucous and cause complications. Within reason the patient should be encouraged to drink freely.

**Exercise and Cystic Fibrosis**

Patients who have cystic fibrosis should be encouraged to exercise. Physical exercise can improve psychological outlook and quality of life, and it may improve pulmonary function, strengthen the respiratory muscles, and decrease bone loss.

**Physical Therapy and Cystic Fibrosis**

Physical therapy has long been one of the mainstays of treatment for cystic fibrosis. The physical therapy techniques used to treat patients who have the disease are very specific and unlike what most people think of when they imagine physical therapy. These techniques have also evolved far beyond the
traditional techniques of postural drainage and chest percussion, and the term
that is commonly used today for physical therapy in cystic fibrosis is airway
clearance techniques. Some of the ones that are commonly used are:

- **Autogenic drainage**: Autogenic drainage can be performed by the
  patient after she/he has been trained to do it. There are various types
  of autogenic drainage techniques but they all utilize a specific pattern
  of timed inhaling, time breath holding, and timed exhaling, all of which
  are slowly increased in duration and/or force. Autogenic drainage
  moves mucous out of the lungs and into the upper airways and oral
  cavity, at which point it can be expelled. The patient is taught to listen
  to his/her breathing and to feel the chest. Doing so will determine
  where the mucous secretions are, and this self-monitoring provides the
  cues for increasing the intensity of the exercise. A full cycle of
  autogenic breathing can take 20-45 minutes.

- **Postural drainage**: Postural drainage is very simple. The patient is
  placed in a position that allows gravity to move mucous from the lungs
  to the upper airways and the oral cavity. For example, the patient may
  be asked to lie on her/his back, tilted down with the head below the
  feet. Postural drainage is often used in conjunction with percussion.

- **Percussion**: Percussion is done by cupping the hand and rhythmically
  hitting the chest over the ribs. The force and vibration that is produced
  will help loosen mucous. When percussion is done by an experienced
therapist the patient will certainly feel the force of the percussion but it will not be painful.

- High frequency chest oscillation: High frequency chest oscillation could be thought of as mechanical chest percussion. The high frequency chest oscillator uses a vest that is attached to an air generator by two hoses. The air generator alternately inflates and deflates the vest, providing much the same effect as manual percussion. The mucous that is loosened from the airways can then be coughed out by the patient.

- Positive expiratory pressure (PEP): Positive expiratory pressure uses a face mask that is similar to an oxygen face mask. The mask has a valve that can be adjusted so that exhaling requires a greater than normal force and the increased work of exhaling, along with periodic coughing, can help remove mucous.

**Medications and Cystic Fibrosis**

Medications cannot cure cystic fibrosis; they are used to manage and to prevent specific complications. For example, patients who have cystic fibrosis often have lung infections and these are treated with antibiotics. (Note: In the module the generic names of drugs are used first and common trade names are capitalized and placed in parentheses)

- Antibiotics: Antibiotics can be given intravenously, orally, or by using a nebulizer. Antibiotics are only used to treat active, ongoing infections; they should not be used chronically as a preventive measure.
• Anti-inflammatory drugs: Some anti-inflammatory drugs such as ibuprofen (Motrin) can be used for certain patients who have cystic fibrosis. However, long-term use of these drugs is not recommended.

• Biphosphonates: The biphosphonates such as alendronate (Fosamax) increase bone density and they can be used if the patient has evidence of bone loss.

• Bronchodilators: Excess mucous, damage to the airway and in some patients narrowing of the airways that is similar to asthma, decrease the size of the bronchial passages and make breathing difficult for the patient who has cystic fibrosis. Bronchodilators such as albuterol (ProAir, Ventolin) and ipratropium (Atrovent) dilate the airways and increase oxygen delivery. These drugs are not typically prescribed for routine use but are used when the patient is having an airway clearance technique performed or when she/he feels especially short of breath.

• Insulin: Cystic fibrosis patients who have diabetes mellitus require insulin.

• Mucolytics: Lysis is a medical term that means to break down. Mucolytics such as hypertonic saline and dornase alfa (Pulmozyme) are administered by a nebulizer; they break down and thin out mucous, making it easier to cough up.

• Oxygen: Supplemental oxygen can be used for patients who do not have normal blood oxygen saturation.
- Pancreatic enzymes: Pancreatic enzymes such as Creon or Pancreaze are considered to be standard care for patients who have cystic fibrosis.
- Vaccines: patients who have cystic fibrosis should be given influenza and pneumococcal vaccines.
- Vitamin supplementation: Vitamins A, D, E, and K are routinely given to patients who have cystic fibrosis.

**Genetic Modulators and Cystic Fibrosis**

The medications that were discussed in the previous section can be effective for easing the severity of the signs and symptoms of cystic fibrosis, but they do not address why the disease happens. However, a relatively new class of drugs called genetic modulators has shown much promise as a treatment for the root cause of cystic fibrosis.

Cystic fibrosis is caused by a defect in the gene that controls how cells transfer the electrolytes chloride and sodium across the cell membrane. Abnormal transfer of these electrolytes makes the mucus thick and sticky, and the genetic modulators such as ivacaftor (Kalydeco) improve the function of this gene. The drugs are taken orally and the genetic modulators have been shown to increase breathing capability, decrease gastrointestinal complications, and decrease the number of respiratory problems.

**Surgery and Cystic Fibrosis**

For some people who have cystic fibrosis, a lung transplant is necessary. If they are producing large amounts of mucus that cannot be managed with
medications, diet, or exercise, and/or if they are having persistent lung infections that do not respond to antibiotics, a lung transplant may be the only treatment that can help. If the patient's gastrointestinal tract is compromised, a feeding tube may need to be surgically placed.

**CARING FOR A PATIENT WHO HAS CYSTIC FIBROSIS**

Caring for a patient who has cystic fibrosis can be very challenging. It can also be very rewarding as well, because your interventions can make a big difference. The patient who has cystic fibrosis needs skilled care and psychological support to successfully cope with this illness and as a health care professional, you can monitor the patient's condition and provide encouragement.

When you are caring for a patient who has cystic fibrosis, focus on these five areas of care.

1. **Infection control:** The patient who has cystic fibrosis is very susceptible to developing respiratory tract infections so adherence to standard precautions is very important. All CNAs are familiar with standard precautions and standard precautions should, of course, be used when caring for any patient. As a review, standard precautions includes: a) hand washing; b) the use of personal protective equipment; c) respiratory and cough etiquette; d) considering all body fluids to be potentially infectious, and e) injection safety. All of these are important but when caring for a patient who has cystic fibrosis hand washing and respiratory and cough etiquette are especially so. Cystic fibrosis is not an infectious disease so these patients do not need to be in isolation. If
you have an infectious disease and especially if you have a respiratory infection, you should not be caring for a patient who has cystic fibrosis.

2. Monitoring for infections: Frequent assessments of temperature, respiratory status (This is discussed in the following paragraph) and the production of mucous are essential. Immediately notify a nurse or your supervisor if the patient has a fever, the respiratory status is abnormal, or there is a change in the patient’s mucous production. If the mucous is green, yellow, or has an unpleasant odor this can be a sign of a respiratory infection.

3. Monitoring respiratory status: Remember, patients who have cystic fibrosis are very susceptible to developing respiratory tract infections, and respiratory tract infections are the major cause of death for these patients. You will need to perform a basic assessment of the patient’s respiratory status to make sure they are breathing normally. Check the respiratory rate, check skin color, look for cyanosis in the nail beds, and measure oxygen saturation if this is required. Most importantly, ask the patient if she/he is short of breath or if his/her breathing feels abnormal. The patient is often the most reliable source for determining respiratory status. The respiratory rate, the skin color, and the oxygen saturation level may all be normal, but if the patient reports that she/he is having difficulty breathing that information is more important than the physical assessment. You should also determine how easy or difficult it is for the patient to cough up mucous.
4. **Hydration and nutrition**: Good hydration and good nutrition are essential for patients who have cystic fibrosis. Dehydration will cause the mucous in the lungs and the gastrointestinal tract to become thicker and more viscous and this can lead to complications. The patient's physician will establish guidelines for how much fluid the patient is allowed to have and within those limits you should encourage the patient to drink. If the patient cannot or will not hydrate, this should be reported to a nurse or your supervisor. Nutritional deficiencies can easily occur in this patient population and the patient should be encouraged to eat. The patient's physician will establish guidelines for nutrient intake. Familiarize yourself with these guidelines, monitor the patient's nutrient intake and report any problems to a nurse or your supervisor.

5. **Monitor for complications**: Patients who have cystic fibrosis are at risk for respiratory and/or gastrointestinal complications. These complications will usually present as infections, difficulty breathing, or gastrointestinal complaints. If the patient has a fever, difficulty breathing, or a problem with his/her stomach of bowels, notify a nurse or your supervisor.

**SUMMARY**

Cystic fibrosis is a genetic disease that causes the production of large amounts of thick, viscous mucous. The excess and excessively thick mucous has many complicated effects on the body, but cystic fibrosis is primarily known as a
disease that affects the respiratory tract. Patients who have cystic fibrosis have bronchial airway passages that are blocked with mucous, preventing the movement of oxygen through the lungs. The mucous is also very irritating to the lungs, and inflammation and damage to the airways is common. Finally, the mucous provides an ideal place for microorganisms to grow and respiratory infections are the most common cause of death in people who cystic fibrosis.

Other common complications of cystic fibrosis include diabetes mellitus; cirrhosis of the liver, gall bladder disease, osteoporosis, and infertility. Diabetes mellitus is perhaps the most serious of these. Approximately 50% of all patients who have cystic fibrosis have diabetes mellitus and the presence of this complication is very serious: diabetes mellitus along with cystic fibrosis increases the risk for respiratory infections and decreases the rate of survival.

Cystic fibrosis can be detected pre-natally and all babies are required to be tested for the disease shortly after birth. However, there is no cure for cystic fibrosis and the outlook for someone who has the disease is very poor. Despite advances in care and the development of dedicated cystic fibrosis care centers many people who have cystic fibrosis will die by age 40.

Treatment for cystic fibrosis is primarily focused on monitoring the patient for complications and treating them as they arise; maintaining good nutritional status, and; monitoring for, preventing, and treating respiratory infections. These therapeutic goals are achieved by the use of:

- Diet: Hydration, a high-calorie diet with extra fat, and supplemental amounts of vitamins A, D, E, and K.
• Exercise: Exercise can increase the strength of the respiratory muscles, improve psychological outlook, improve pulmonary function, and decrease bone loss.

• Physical therapy: Physical therapy techniques for the treatment of cystic fibrosis are called airway clearance techniques. These therapeutic modalities are designed to move excess mucous from the lower parts of the lungs to the trachea and the oral cavity, at which point they can be expelled by coughing. Many of the airway clearance techniques are used together and they can also be used with bronchodilators.

• Medications: The majority of the medications used to treat cystic fibrosis provide symptomatic relief that addresses the complications of the disease: Antibiotics, anti-inflammatories, bronchodilators, mucolytics, and oxygen for the respiratory issues; pancreatic enzymes to help digestion; bisphosphonates for bone loss, and; vaccines to prevent influenza and pneumonia. The genetic modulators improve the function of the defective gene that causes cystic fibrosis, treating the disease, not its consequences.

• Surgery: Lung transplantation is an option for patients who are having persistent complications and do not respond to standard therapy. Some patients who have nutritional deficiencies but cannot eat may require placement of a feeding tube.
The responsibilities of a CNA when caring for a patient who has cystic include close attention to infection control; monitoring for respiratory complications; monitoring the patient’s respiratory status, and; encouraging the patient to eat and drink and ensuring that she/he is well hydrated and well nourished. Specific tasks during the day would include:

- Assessing skin color
- Assisting the patient to exercise
- Documenting food intake
- Examination/assessment of the mucous production
- Measuring body temperature
- Measuring oxygen saturation
- Measuring pulse and respiratory rate
- Preparing the patient for airway clearance techniques
- Recording intake and output