UNDERSTANDING CYSTIC FIBROSIS

Abstract:

Cystic fibrosis is a genetic disease that primarily affects the lungs, but may also have serious adverse effects on many other organ systems. In cystic fibrosis the genetic information is faulty and causes a serious condition to develop where thick, viscous secretions are produced that affect the lungs, gastrointestinal tract, including the pancreas, and other body organs. There is no cure for cystic fibrosis but medications, physical therapy, diet, and lifestyle adjustments can help prevent complications by making life easier for patients to live with the disease.

Learning Objectives:

1. State the cause of cystic fibrosis.
2. Identify the signs and symptoms of cystic fibrosis.
3. Describe the treatment options and the basic care needed for someone diagnosed with cystic fibrosis.

Introduction

Cystic fibrosis is a serious, chronic disease that affects the respiratory system, gastrointestinal system, pancreas, and many other organs. People with cystic fibrosis have a genetic defect that causes production of an excess amount of thick viscous mucus that damages specific organ systems, causing serious respiratory, metabolic, and gastrointestinal complications. There is no cure for cystic fibrosis. 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 lower than the average, however, medications, physical therapy, diet, and lifestyle adjustments can help prevent complications and improve patient quality of life.

Causes and Risks of Cystic Fibrosis

The tendency to develop cystic fibrosis is inherited. In genetically inherited conditions the genes are found in pairs and determine a cells structure and how it functions. Genes are the part of a cell that contains the information that regulates how cells reproduce. Cystic fibrosis is an autosomal recessive disorder. This means a patient must have two of these abnormal genes, one from each parent. Cystic fibrosis will present if both of the genes that control the production of mucus are not normal.

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. The gene involved in cystic fibrosis is responsible for the production of mucus in the lungs, gastrointestinal tract, pancreas and other areas. In cystic

fibrosis, the genes involved control the cells that produce mucus but the genetic information is faulty and the mucus that is produced is viscous, thick and very sticky.

Mucus 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. Mucus also traps bacteria and viruses and prevents them from causing infections and in parts of the gastrointestinal tract mucus helps digest food and helps move digested food through the gut. For people without cystic fibrosis, mucus is relatively thin and watery. In patients with cystic fibrosis, the gene that regulates mucus production is abnormal and people who have this disease produce mucus that tends to adhere to the walls of the lungs and the gut. The abnormal mucus can block the bronchi and other air passages in the lungs. It can also block the gastrointestinal tract in various places such as the pancreas, gallbladder, and bowels. This thick, sticky mucus 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.

A person who inherits a copy of the defective gene that causes cystic fibrosis from each parent will develop cystic fibrosis. A person who inherits only one copy of this gene will not.

Cystic fibrosis is the most common, lethal inherited disease in the white population. In the United States, cystic fibrosis affects approximately 1 of 3,500 Caucasians, 1 of 9,500 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 cases the disease develops during adulthood.

Cystic Fibrosis Symptoms and Complications

Cystic fibrosis is a disease that primarily develops in the lungs. 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 a primary concern. As mentioned, the mucus in the lungs of people who have cystic fibrosis is thick, viscous, and sticky. There is also an abnormally large amount of mucus produced. These factors have many severe implications.

In the normal lungs, mucus gets removed in several ways. Coughing and sneezing help expel some mucus, and move a large amount of mucus to the trachea and the oral cavity, at which point it can be expelled or swallowed. Mucus 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 mucus in the respiratory passages up and out towards the trachea and the mouth. At that point the mucus can be removed by involuntary and voluntary

coughing, sneezing or swallowed. This process is called the mucus escalator.

Because the mucus in the lungs of someone with cystic fibrosis is dense and thick, it adheres tightly to the airway passages, and there is a large amount of mucus being produced. Coughing, sneezing, and the mucociliary escalator are not able to remove the mucus effectively, and excess mucus stays trapped in the lungs.

The terms mucus, phlegm, and sputum are often used interchangeably when discussing cystic fibrosis. Phlegm is abnormally thick mucus produced in the respiratory tract, and sputum is mucus 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 mucus that is produced harms the lungs in several ways.

The mucus forms a barrier, plugging the small and medium-sized airways and preventing oxygen from moving from the lungs into the bloodstream. Irritation and inflammation of the small and medium-sized airways is caused by mucus in the airways, eventually damaging the airways.

The presence of mucus in the airways provides an ideal environment for bacteria to grow. Bacterial lung infections are common in people who have cystic fibrosis. The great majority of people who die from cystic fibrosis succumb because of 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 mucus production prevents oxygen from

moving through the lungs, so people who have cystic fibrosis often feel short of breath. The excessive, thick mucus irritates 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

Diabetes mellitus is a common complication of cystic fibrosis. There are three ways that cystic fibrosis causes diabetes: pancreatic damage, insulin release, and insulin resistance.

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 mucus 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.

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.

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, and they must be treated with insulin.

Digestive Diseases

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. First, the excess, thick mucus in the digestive tract can block the bowels, the bile ducts

of the gallbladder, and the drainage ducts of the pancreas. Second, the mucus provides an environment that allows bacteria to grow and the gastrointestinal (GI) tract and its organs can become inflamed. Because of these pathologic processes, people who have cystic fibrosis are prone to various diseases of the gallbladder, liver and GI tract, as well as complications related to malabsorption.

Gallbladder and Nutritional Deficiencies:

Bile is a compound that is produced in the liver and stored in the gallbladder. Bile is secreted from the gallbladder 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 mucus 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 gallstones and gallbladder infection.

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 GI tract at the time of birth and meconium is very thick and tenacious. Bowel obstructions can also occur in other parts of the GI tract in adults.

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. Dietary sources are relied upon for calcium intake 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.

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 gallbladder become obstructed with mucus and become inflamed.

Reproduction

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 multifactorial.

Other Complications

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

Table 1: Signs and Symptoms of Cystic Fibrosis

Abdominal pain
Constipation
Cough
Decrease exercise tolerance
Diarrhea
Dry skin
Dyspnea
Fatigue
Flatulence
Hyperglycemia
Tachypnea
Weight loss
Wheezing

Diagnosing Cystic Fibrosis

Cystic fibrosis is usually diagnosed by age 6-8 months. It is diagnosed using the three methods of genetic testing, sweat test, and respiratory and pancreatic complications.

Genetic Testing

Genetic testing to detect cystic fibrosis can be done in the prenatal period or postnatally. The prenatal genetic screening tests are very sensitive and 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 Chloride Test

People with cystic fibrosis produce sweat that has a high sodium (salt) content because people with cystic fibrosis do not reabsorb salt from

their sweat as they should. To perform the test, a chemical is used to make the skin sweat when triggered by a weak electric current. The sweat is then collected to analyze. If the initial test shows high sodium content in the sweat, the test is repeated several weeks later for confirmation. If the sweat test is negative but a 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.

The sweat chloride test is the most common, simple test used to diagnose cystic fibrosis. Other tests used are briefly listed below.

Immunoreactive Trypsinogen (IRT) Test

In newborns, the immunoreactive trypsinogen test a standard screening tool used to measure whether abnormal levels of the protein immunoreactive trypsinogen exists in the newborn's blood. When there is a high level of IRT, this could indicate the presence of cystic fibrosis. However, supplementary testing is necessary to verify this diagnosis.

Pulmonary Function Tests (PFTs)

Pulmonary function tests (PFTs) are performed to determine whether a person's lungs are properly functioning. The test measures the amount of air inhaled and exhaled and the level of oxygen is transferred throughout the rest of a person's body. Abnormalities that arise during testing may indicate the presence of cystic fibrosis.

CT Scan and Chest X-Ray

A computed tomography (CT) scan is done when detailed imaging of a person's internal organs is needed. The CT scan is performed through the combination of x-ray imaging, and these scans are able to show the extent of liver or pancreas damage indicative of cystic fibrosis.

A chest X-ray may also be done that will reveal swelling in the lungs due to obstructions in the respiratory passageways from the thick, viscous secretions that occur in a case of cystic fibrosis.

Sputum Test

The presence of a lung infection may also be determined by a sputum test, where the medical clinician obtains a small sample of mucus. Patients with cystic fibrosis are prone to pulmonary infections, and the mucus sample is used to determine bacteria that are present, which helps guide the clinician to determine the best treatment to treat infection if it should exist.

Prognosis for Cystic Fibrosis

There is no cure for cystic fibrosis and unfortunately a diagnosis of cystic fibrosis is accompanied by a 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 for the great majority of people, cystic fibrosis is serious.

The median life expectancy of a person diagnosed with cystic fibrosis is approximately 40 years and 20% of people who have cystic fibrosis do not live to adulthood. Advances in treatments have improved the

outlook for these patients and now more than half the people with cystic fibrosis are 18 years or older. The median life expectancy (the middle number in a sequence or population) of 40 years of age 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.

Factors that contribute to a poor prognosis are diagnosis at a young age, severe symptoms, rapid progression of the disease, the presence of diabetes mellitus, and the complexity of the disease. Some patients who have cystic fibrosis may survive into their 50s and this is a far better outlook than what was historically expected. 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

Treatment of cystic fibrosis involves diet, exercise, medications, physical therapy and occasionally, surgery. The treatment of this disease 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. The treatment of cystic fibrosis also requires intense involvement of the patient and the patient's family.

Living with cystic fibrosis is difficult, and, although there is no cure for the disease, there are three main goals of treating cystic fibrosis that can help improve the quality of a patient's life. These goals include 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

Consultation and periodic reviews with a nutritionist are 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, a patient who has cystic fibrosis does not need a highly specialized or restrictive diet unless diagnosed with diabetes.

Good hydration is also important because dehydration can increase the thickness of mucus and cause complications. Within reason the patient should be encouraged to drink freely.

Exercise

Patients who have cystic fibrosis should be encouraged to exercise because sweat glands and airway cells are activated when exercising. Physical exercise may also refine psychological outlook regarding the quality of life by decreasing bone loss, increasing cardiovascular fitness, and facilitating mucus clearance to improve pulmonary function and strengthen the respiratory muscles.

Physical Therapy

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 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 discussed next.

Autogenic Drainage:

Autogenic drainage can be performed by a patient after being trained to do it. There are various types of autogenic drainage techniques but they all utilize a specific pattern of timed inhaling, timed breath holding, and timed exhaling, all of which are slowly increased in duration or force.

Autogenic drainage moves mucus out of the lungs and into the upper airways and oral cavity, at which point it can be expelled. Patients are taught to listen to their breathing and to feel the chest. Doing so will determine where the mucus 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 simple. The patient is placed in a position that allows gravity to move mucus from the lungs to the upper airways and the oral cavity. For example, a patient may be asked to lie on the 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 mucus. 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 mucus 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 mucus.

Medications

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. The medications discussed below will list the drug generic names first and the common trade names 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 frequently 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 a patient has evidence of bone loss.
Bronchodilators:

Excess mucus, 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 a 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 a patient is having an airway clearance technique performed or when feeling 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 mucus, 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

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 GI complications, and decrease the number of respiratory problems.

Surgery

Most present-day medical management strategies are able to control cystic fibrosis complications well into adulthood. However, the lungs may continue to produce large quantities of mucus that cannot be managed with medications, diet, or exercise. Therefore, a lung transplant contributes to prolonged survival with this type of advancing pulmonary disease.

Caring for Patients with Cystic Fibrosis

A patient who has cystic fibrosis needs skilled care and psychological support to successfully cope with this illness. As a member of the healthcare team, a CNA may monitor a patient's condition and provide encouragement. There are five areas of care a CNA should focus on when caring for a patient who has cystic fibrosis. These include infection control, monitoring for infections, monitoring respiratory

status, monitoring for complications, and maintaining hydration and nutrition.

Infection Control

A patient who has cystic fibrosis is susceptible to developing respiratory tract infections so adherence to standard precautions is important. All CNAs should be familiar with standard precautions and standard precautions are to be used when caring for any patient. As a review, standard precautions includes 1) hand washing, 2) the use of personal protective equipment, 3) respiratory and cough etiquette, 4) considering all body fluids to be potentially infectious, and 5) 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 important.

Cystic fibrosis is not an infectious disease so these patients do not need to be in isolation. A CNA who has an infectious disease, especially a respiratory infection, should not be caring for a patient who has cystic fibrosis.

Monitoring for Infections

Frequent assessments of temperature, respiratory status and the production of mucus are essential. The CNA should immediately notify the supervising nurse or physician if a patient has a fever, an abnormal respiratory status, or a change in mucus production. If the mucus is green, yellow, or has an unpleasant odor this can be a sign of a respiratory infection.

Monitoring Respiratory Status

The CNA needs to remember that 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. The CNA will need to perform a basic assessment of the patient's respiratory status to make sure that breathing is normal. The CNA should check the respiratory rate, skin color, nail beds (for cyanosis or blue color indicating low oxygenation), and measure oxygen saturation if this is required. Most importantly, the CNA should ask the patient if he or she is short of breath or if 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 a patient reports breathing feels difficult then that information is more important than the physical assessment. A CNA should also determine how easy or difficult it is for a patient to cough up mucus.

Monitoring for Complications

Patients who have cystic fibrosis are at risk for respiratory or GI complications. These complications will usually present as infections, difficulty breathing, or gastrointestinal complaints. If a patient has a fever, difficulty breathing, or a stomach or bowel problem, the CNA should notify the supervising nurse or physician.

Specific patient care tasks that a CNA may perform during the day are highlighted in Table 2 below.

Table 2: Patient Care Tasks

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

Hydration and Nutrition

Good hydration and nutrition are essential for patients who have cystic fibrosis. Dehydration will cause the mucus in the lungs and the GI tract to become thicker and more viscous and this can lead to complications. A patient's physician will establish guidelines for how much fluid the patient is allowed to have and within those limits the CNA should encourage the patient to drink. If a patient cannot or will not hydrate, the CNA should report this to the supervising nurse or physician. Nutritional deficiencies can easily occur in this patient population and the patient should be encouraged to eat.

A patient's physician will establish guidelines for nutrient intake. The CNA should be familiar with these guidelines, monitor the patient's nutrient intake and report any problems found.

Case Study

The following is a case study of a 9-month year old female diagnosed with cystic fibrosis shortly after birth.

The patient was diagnosed at 4 months of age with cystic fibrosis and also with pseudomonas lung infection. She was hospitalized in the pediatric intensive care unit (PICU) for 3 months with respiratory failure that required emergency intubation. She eventually recovered from this episode after responding well to treatment and was able to discharge home for a brief while. Six months later, she was readmitted to the hospital with vomiting induced by coughing that had been lasting for about a week before her parents brought her to the hospital emergency room. Her mother had noticed the patient had difficulty breathing and had to work harder at breathing especially after a coughing episode. The patient also had a poor appetite for 4 days prior to admission, and showed symptoms of increased sweating and decreased wet diapers. On this admission, the patient required another PICU stay for severe metabolic insufficiency (low blood sodium and low blood potassium).

Discussion

Severe low blood sodiums with weight loss are often a symptom that parents may bring their child into hospital for prior to having an understanding of what is causing the condition. Its not unusual for parents of young infants to bring them to the hospital because of weight loss due to an unknown cause and with severe dehydration.

In such clinical situations the child can improve with sodium chloride added to their feedings. When clinical symptoms continue to worsen, such as weight decreasing regardless of nutrient intake, the clinician may suspect that cystic fibrosis is a cause. The sweat test can be done as well as a full gene sequencing to rule out cystic fibrosis. Once positively diagnosed, the child's body organs, such as the lungs and GI

tract, may be evaluated for disease. The clinician will also look for multiple complications common to cystic fibrosis patients.

The child's weight will be monitored to determine failure to thrive in a suspected or confirmed case of cystic fibrosis. Diet is assessed, and the questions a clinician may ask the parents would pertain to whether their child is provided formula feeds. In the case of the patient above, at the time of her first admission her parents reported she was breastfed and the pediatrician recommended Similac Advance every 3 hours (7-8 feeds per day). Supplements were added to the patient's diet, which included 1/8 tsp salt daily.

The pediatrician recommended that the parents continue Similac Advance feedings every three hours as able with a minimum goal of feeding to support growth. The parents were advised to continue with supplements that included pancreatic enzymes in addition to salt 1/8 tsp daily. They were recommended to ensure the vitamins, enzymes and salt were administered daily and that the patient's daily weights were being monitored with a goal to catch-up with growth. The parents were also asked to monitor the patient's stools.

By the time the patient approached 1 year of life, she had good oral intake and was having 2 and sometimes 3 stools daily. Her growth trends improved and she exceeded the max goal of dietary intake whereas previously her appetite was low. This case helps to illustrate the benefit of basic cystic fibrosis treatment in an infant girl during the first year of life and the difference to her growth and development with basic dietary and supplementation treatment. Care of the adult would be different, and the basic treatment for a adult, depending on the

adults level of disease and cystic fibrosis complications would determine the course of care.

Summary

Cystic fibrosis is a genetic disease that causes the production of large amounts of thick, viscous mucus, and is primarily known as a disease that affects the respiratory tract. Patients who have cystic fibrosis have bronchial airway passages that are blocked with mucus, preventing the movement of oxygen through the lungs. The mucus is also irritating to the lungs, and inflammation and damage to the airways is common.

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, gallbladder disease, osteoporosis, and infertility. Cystic fibrosis can be detected prenatally and all babies are required to be tested for the disease shortly after birth. There is no cure for cystic fibrosis and the outlook for someone who has the disease is poor.

Treatment for cystic fibrosis is primarily focused on monitoring the patient for complications and treating them as they arise, and maintaining good nutritional status. Also, health team members must monitor for the existence of a respiratory infection and educate the patient on ways to prevent an infection from happening. These therapeutic goals are achieved by the use of diet, exercise, physical therapy, medications, genetic modulators (improving the function of the defective gene causing cystic fibrosis), and surgery.

The responsibilities of a CNA when caring for a patient who has cystic fibrosis 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 to ensure the patient remains well-hydrated and nourished. Although there is no cure for cystic fibrosis, the use of medications, physical therapy, diet, and lifestyle adjustments can help prevent complications and improve the patients quality of life in all age groups.