Cystic Fibrosis: A Pediatric Case Study

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Introduction

During my four years obtaining a Bachelor of Science in Nutrition from Drexel University, I participated in and helped organize many fundraisers for the Cystic Fibrosis Foundation (CFF). Now, as I advance my career as a healthcare professional, I am intrigued by the nutritional complications that often occur with this disease. Cystic fibrosis (CF) is primarily considered to be a pulmonary disease, however there are many other organs and metabolic functions that are affected. These pediatric patients require a large interdisciplinary team effort to deliver appropriate care, including Registered Dietitians, Nurses, Pulmonologists, Respiratory Therapists, Social Workers, and often Gastroenterologists. Registered Dietitians have the large responsibility of helping CF patients maintain adequate growth and weight gain from infancy to adulthood. Due to malabsorption issues, the task of maintaining appropriate growth can be very difficult at times, and malnutrition is common in this disease population.

While technology and medicine are becoming more advanced, CF patients are living longer and longer, past childhood and adolescent years. Although the predicted life expectancy age is increasing, it is still critical for each patient to keep up with countless treatments every single day. Family support may be crucial to urge young children to do treatments in order to have the best possible outcomes with this disease. Upon researching this case study, I sought after learning the nutritional, medical, and emotional struggles that pediatric CF patients experience throughout childhood, and the methods currently being used to overcome these struggles.
Abstract

Cystic fibrosis (CF) is an inherited autosomal recessive chronic disease that causes the body to produce thick and sticky secretions. The mucus secretions primarily affect the lungs, as well as the pancreas, and other exocrine glands in the body. The CF gene needs to be inherited from both parents in order for a child to have the disease. The predicted median age of a person living with CF is in the early forties.

Medical nutrition therapy for cystic fibrosis can be complicated and involves monitoring macronutrients, as well as many micronutrients. Sodium chloride repletion is needed due to excessive losses through the skin. Due to frequent malabsorption issues in the intestines, vitamin and mineral deficiencies are common, especially fat-soluble vitamins due to fat malabsorption.

My patient, RW, is a 9-year-old Caucasian male with CF, who was diagnosed around 7 months old. Some of his medical history included failure to thrive as an infant, pancreatic insufficiency, asthma, gastrointestinal reflux, malnutrition, and a previous gastrostomy tube placement with subsequent removal. He comes from a complicated family background and has recently been facing signs of depression. On this hospital admission, medical management consisted of the interdisciplinary team agreeing to the patient’s request to replace a percutaneous endoscopic gastrostomy tube for tube feeding.

The nutrition assessment determined RW needs at least 2100 calories per day. He will be receiving a nocturnal tube feeding each night to meet 87% of his estimated nutrition needs. An elemental formula, Elecare Junior, will be used in order to assist with better absorption and requires no need for pancreatic enzymes. Pancreatic enzymes will still be taken with meals and snacks during the day, however, the nocturnal tube feeding provides RW with more freedom to eat how he pleases during the day to be a “normal kid” instead of taking supplements at school.
Discussion of Medical Condition

What is Cystic Fibrosis?

Cystic fibrosis (CF) is an inherited chronic disease that causes the body to produce unusually thick and sticky secretions. The mucus and secretions cause buildup that primarily affects the lungs and pancreas, as well as the gastrointestinal (GI) tract, bile ducts and liver, sweat glands, and other exocrine glands in the body. This disease is most commonly characterized by recurrent lung infections since the mucus buildup attracts bacteria. In newborn infants, meconium ileus is a classic sign of CF, which is sticky mucus and stool accumulation that builds up in utero. Some other symptoms include high levels of sodium chloride in saliva, tears, and sweat, or “salty” skin, persistent coughing with wheezing, shortness of breath, and phlegm, poor growth and weight gain as a child, and greasy, bulky stools.

Most cases of CF are diagnosed at birth and more than 70% of the cases are diagnosed by age 2. One thousand new cases are diagnosed each year. CF affects about 30,000 Americans, and 70,000 people worldwide. The disease is most common among Caucasians and about one in every 3,200 Caucasians are affected.

In the past, CF had typically been associated with younger children, and they would barely live into their teenage years. Due to many breakthroughs in medicine and technology in the past couple decades, CF patients are living to be older and older. Currently, more than 45% of the CF patient population is age 18 or older, and the expected median age of survival for a person living with CF is now the early 40s. Some children also refer to the disease as “65 roses” because it is much easier to pronounce than cystic fibrosis.

Genetics of Cystic Fibrosis

Cystic fibrosis is an autosomal recessive disease that children must inherit from both parents. More than 10 million Americans (or 1 in 31 Americans) are carriers of the CF gene.
and are not aware that they carry it. Also, about 2-5% of all Caucasians carry the CF gene. CF is much less common among other races, about 1 in 46 Hispanic Americans, 1 in 65 African Americans, and 1 in 90 Asian Americans are carriers of the CF gene. In order for parents to determine whether they are carriers and could pass the gene on to their future children, they may undergo genetic carrier testing. Genetic carrier testing can be done for a number of diseases that children may inherit from their parents, one of them being cystic fibrosis.

The cystic fibrosis transmembrane regulator (CFTR) is the gene on chromosome 7 that is responsible for regulating the flow of sodium and chloride ions in and out of cells across the cell membrane. When there are mutations made to this gene, the flow of ions is irregular and adversely affects multiple organs in the body. Since 1989, about 1,800 mutations of the CF gene have been found, but not all genetic mutations will cause the disease. Some common mutations known to cause cystic fibrosis include nonsense or stop mutations (G542X), gating mutations (G551D), protein processing mutations (Delta F508), and other mutations (R117H). Knowing the type of mutation that a child has inherited may help guide the care and any potential new drugs the child could receive in the future.

Results of genetic carrier testing will determine if either, or both parents are carriers of the gene. If both parents are carriers of the CF gene, there is a 1-in-4 chance that the child will have CF, a 2-in-4 chance that the child will also be a carrier of the CF gene, and a 1-in-4 chance that the child will neither have CF nor be a carrier of the CF gene. Carriers of the CF gene do not have the disease or any of the symptoms. If parents do not have genetic testing done before or during pregnancy, then babies are tested at birth.

**Newborn Screenings and Diagnosis of CF**

The most common way to assess newborns for diseases is through newborn screening (NBS) done at birth. NBS involves taking a few drops of blood from the newborn’s heel,
mailing it to a special laboratory to be tested for up to 50 diseases and conditions, depending on the specific state the child is born in. The reasoning behind these newborn tests is that the earlier most diseases are diagnosed, the better doctors can treat your baby to keep him/her as healthy as possible. Results are usually sent to your pediatrician or primary health care provider within about one week. If the NBS comes back positive or abnormal, then diagnostic tests will need to be done to determine if your baby has a health condition or not. Most babies with a positive newborn screen for CF do not end up having the disease. The CF screening test, part of the NBS, is screening for a chemical from the pancreas, called immunoreactive trypsinogen (IRT). While this chemical is normal to find in the body, it tends to be higher in babies with CF. There are other reasons that IRT could be high as well, such as a premature birth or stressful delivery for example, which is why another test needs to be done on these babies in order to determine if the baby actually has CF. Ninety percent of newborns with the high IRT level from NBS have a second screening to check the gene that causes CF.

Babies with a positive newborn screen for CF, will then need to undergo a “sweat test” in order to diagnose the disease. The sweat test is the “gold standard” for diagnosing babies with CF and will measure the amount of salt in the baby’s sweat, taken from the arms and/or legs. During the sweat test, a gel is placed on the skin of an arm or leg, and small wires with patches are placed on top of the gel, and this will cause the skin to feel warm and sweat. Sweat will then be collected for 30 minutes with a gauze pad and sent to a laboratory in order to be analyzed for sodium and chloride (salt) content. A high amount of salt (chloride values > 60 mEq/L) means that the baby has CF, and a low amount of salt means the baby does not have CF. If the results are inconclusive, then a second test will need to be done if necessary. Gene analysis and sputum cultures may also be needed in order to diagnose CF. Children who are diagnosed earlier in life tend to be healthier and have better nutrition than those diagnosed later.
Weight and Nutritional Status of CF Patients

Weight status of patients does have an affect on outcomes in this disease. There is a link between the extent of malnutrition and exacerbation of CF symptoms\(^1\). Maintaining good levels of nutrition can help to increase the long-term survival. Diagnosing early can help with an aggressive nutritional approach for better results. The main goals with these patients are to avoid developing malnutrition, and promote growth despite malabsorption in the intestines, inadequate food intake with some patients, and increased macro- and micronutrient requirements. Generally patients who receive optimal nutrition will have better pulmonary function than those who are malnourished or receive poor nutrition\(^1\). A body mass index (BMI) at or above the 50\(^{th}\) percentile on the growth chart has been associated with better outcomes in children\(^2\). The Cystic Fibrosis Foundation recommends a BMI of 22 for women and 23 for men as a nutrition goal in the adult population\(^2\). The Academy of Nutrition & Dietetics, previously American Dietetic Association (ADA), recommends that patients with CF attend a minimum of four medical nutrition therapy sessions\(^1\), however it is best to have a nutrition assessment completed yearly\(^2\).

Complications of CF

There are numerous widespread complications that arise with cystic fibrosis due to mucus and secretion buildup that can essentially occur anywhere in the body. Some of the many complications associated with this disease include gastroesophageal reflux disease (GERD), CF-related diabetes (CFRD) and pancreatitis due to pancreatic insufficiency, intestinal and digestive abnormalities, and decreased bone density.

Gastroesophageal reflux (GERD) is also known as heartburn, and results from stomach acid backing up into the esophagus. Different studies state that anywhere from 25% to 100% of CF patients deal with this specific complication\(^2\). Excessive coughing can also make this
complication worse. There are multiple medications that can be used to treat this symptom, as well as surgical procedures to address the issue if it is an ongoing problem.

Due to different issues with the pancreas in CF, one of the complications can be CF-related diabetes (CFRD). Approximately 5-15% of CF patients will develop CFRD, and this number is expected to increase with longer life expectancy. Usually, diabetes would be developed at a later age and suggests that the beta cells are not fully functioning. Among adults with CF, about 40% have some glucose intolerance. Developing CFRD commonly is associated with a decline in nutritional status and overall health.

Pancreatic insufficiency (PI) is one of the most common complications of cystic fibrosis patients. Nearly all CF patients, 85-90%, are pancreatic insufficient. Due to the deficiency of pancreatic enzymes, symptoms of PI include diarrhea, greasy stools, malabsorption, weight loss or poor weight gain, poor growth, vitamin and mineral deficiencies, increased gas, abdominal pain, and essential fatty acid deficiency. With pancreatic enzyme therapy, many of the consequences of pancreatic insufficiency can be slowed or reversed; however, 85% of CF patients show growth retardation. Pancreatitis can also occur due to thick secretions blocking the pancreatic duct, leading to an inflamed pancreas.

Decreased bone density is seen in CF patients, and osteoporosis is now being seen in older CF patients. Some reasons for low bone mass and increased risk for fractures could be due to malnutrition and malabsorption linked to low levels of vitamin D for bone health, inadequate calcium intake, inactivity, delayed puberty, short bowel syndrome, and from medication use, such as corticosteroids. If children have any risk factors for decreased bone density, then a dual energy X-ray absorptiometry (DXA) scan should be done on any patients 8 years of age or older to assess their bone mass status.
Thick liver secretions can lead to decreased bile flow, hepatocyte damage, inflammation, fibrosis, and eventual cirrhosis\textsuperscript{2,3}. In males, infertility may occur if the vas deferens becomes obstructed. Some other common problems include bicarbonate deficiency, abnormalities of bile salts, mucosal transport and motility, and anatomical structural changes of the intestine\textsuperscript{4}. There are many complications that can occur in cystic fibrosis that vary from patient to patient. Thick secretions could build up anywhere in the body and potentially lead to new complications over time.

**Medical Management of CF and Treatments**

Depending on complications and severity of the disease, each individual case of cystic fibrosis is treated a bit differently. Typically, all CF patients go through the same breathing treatments and medications, and most take pancreatic enzymes starting at a young age. Due to malabsorption and excessive loss of sodium and chloride through sweat glands, many vitamins and minerals need to be monitored and replaced as part of overall medical management.

Some specific treatment methods or options include pancreatic enzyme replacement therapy (PERT), airway clearance techniques (ACTs), lung transplantation, pharmacotherapy, gene therapy, and more\textsuperscript{1}.

In CF, the lungs require a lot of attention. There are many treatments and techniques called airway clearance techniques (ACTs) to help reduce the amount of mucus that builds up in the lungs. Since the mucus is thick and sticky, the methods need to be more intense than people without CF who can just cough very hard to bring up mucus. Some ACTs include manual chest physiotherapy (CPT), which is clapping on the patient’s chest and back with your hands, and also high frequency chest compression (HFCC) is another option that includes a vibrating vest hooked up to an air compressor. During HFCC, inhaled medications and breathing treatments can be done at the same time, which is convenient for CF patients who need to make time for all
of their treatments and medications every single day of their life. There are active-cycle breathing techniques (ACBT) to help with breathing control and chest expansion, and they can be done alone if someone is not around to help with a different treatment. ACBTs also include Huff coughing to help force mucus from the smaller airways to the larger airways and coughing it out. The “acapella” is another method that is handheld and provides vibratory positive expiratory pressure (PEP). There are also nebulizer treatments that CF patients can inhale to reduce any bacteria that could be in the lungs, as well as inhalers and medications for asthma.

Lung transplantation is another option for CF patients as they become severely compromised due to their illness and are considered to be in end-stage lung disease. A large obstacle with transplantation is the risk of rejection. There is no weight criterion for children who need a lung transplant, but for adults the weight criteria is 80% to 130% IBW or BMI 18.5 to 30 kg/m\(^2\). This remains the most aggressive therapy for severely reduced lung function in CF patients.

Pancreatic enzyme replacement therapy (PERT) is needed for pancreatic insufficiency. The enzymes given to CF patients include lipase, protease, and amylase to help digest fat, protein, and carbohydrates, respectively. Dosing of enzymes depends on age and individual response to PERT. It is recommended to give 1,000 lipase units per kilogram body weight per meal for patients under 4 years of age, and it is recommended to give 500 lipase units per kilogram body weight per meal for patients over 4 years of age. Dosing for snacks should be cut about half of the meal dose. All doses are based on 3 meals and 2 snacks per day, and should not exceed a dose of 2,500 lipase units per kilogram body weight per meal or 10,000 lipase units per kilogram body weight per day. If patients present with diarrhea or constipation, other gastrointestinal symptoms, or have an abnormal growth pattern, then doses may need to be adjusted. PERT is needed for tube feedings when using a formula with the nutrients fully intact.
PERT may not be needed for elemental formulas that are easier for the gut to absorb. In addition to PERT, pharmacotherapy may be used in combination in order to help balance the acidity of the intestines. Balancing the pH of the gut also helps to achieve near-normal absorption\(^1\).

Gene therapy involves compacting DNA to try to get healthy genes into CF cells; however, more research is being done on this method.

There are CF Foundation-accredited care centers for CF patients to continue care throughout pediatric and adult life. In order to stay healthy and maintain this disease throughout life, CF patients need to continually monitor the diet, stay away from germs, and keep up with their lung health and treatments\(^4\).

**Ongoing Research and Studies**

About 30 prospective drugs are now in development and clinical trials need to be conducted to test if they are beneficial in the CF population\(^4\). Some recent research suggests that sodium nitrite is effective in killing many of the pathogens responsible for causing lung infections in CF, including *Pseudomonas aeruginosa*, *Staphylococcus aureus*, and *Burkholderia cepacia*\(^5\), while other research explains that the distribution of microbial communities throughout the lung is variable depending on location in the lung, which can make infections harder to treat or cure\(^6\).

Every day cystic fibrosis is coming closer to a cure due to the constant efforts and research by many organizations, including the Cystic Fibrosis Foundation (CFF) and Cystic Fibrosis Research Inc. (CFRI). Scientists continue to look for a method to mend the defective CFTR protein since CF gene therapy research was first launched in 1993. Other researchers are scouring over current therapies to determine the best methods of treatment, and the possibility of drug development for a future cure\(^4\).
**Discussion of Medical Nutrition Therapy**

The nutrition assessment is a very critical part of the interdisciplinary care that cystic fibrosis patients receive. Due to many factors affecting nutrition status, such as malabsorption of macronutrients and fat-soluble vitamins, increased oxidative stress, CFRD, and pancreatic insufficiency, all nutrient needs are somewhat individualized. Fluid needs in the CF patient should remain the same as the rest of the population, unless otherwise indicated\(^1\).

**Macronutrients**

There are many methods to calculate caloric needs for pediatric patients, and varying opinions about what is appropriate for CF patients. Each patient will have different needs based on current nutritional status, level of physical activity, fat stores, degree of malabsorption, growth pattern, and the severity of CF\(^2\). One way to calculate calories would be the “Recommended Dietary Allowance” (RDA) recommendations suggesting an amount of calories per kilogram of body weight, based on different ages and gender\(^7\). Two other methods are the World Health Organization (WHO) and Schofield equations to estimate resting energy expenditure (REE). There are different equations for genders, based on ages 0-3 years, 3-10 years, and 10-18 years old. The result is then adjusted with an activity and/or stress factor to better more accurately estimate caloric needs based anywhere from bed rest with mild stress to an active child requiring catch-up growth with severe stress\(^7\). In CF, caloric needs should be calculated for the healthy population using the age, weight, and gender of the patient, and then should be adjusted by 110-200% of the caloric needs\(^2,3\). Using the range of 110-200% of normal caloric needs has shown improved weight status\(^2\). Some sources recommend a smaller range of 120-150% more calories than healthy controls\(^1\). For some teenagers, caloric needs could be reaching around 4000 calories per day. Again, needs are specific to each patient and not one method will apply to all CF patients. Another method states about 150 calories per kilogram of
body weight for children and 200 calories per kilogram of body weight for infants; however, this method seems to estimate very high calories.1

Protein needs will also need to be individualized for each patient. A liberal range of 10-35% calories from protein is recommended for patients with CF.1 Other sources use a more narrow range to recommend 15-20% calories from protein.2 A method used to calculate protein needs as children go through different life stages is estimating 4 grams of protein per kilogram (kg) of body weight for infants, 3 grams of protein per kg of body weight for children, 2 grams of protein per kg of body weight for teenagers, and 1.5 grams of protein per kg of body weight for adults.1 The “Recommended Dietary Allowance” (RDA) recommendations can also be used to calculate protein needs. The RDAs suggest an amount of protein in grams per kilogram of body weight, based on different ages and gender.7 Protein can also be calculated based on level of stress of the body, depending if there were any recent surgeries, any wounds or incisions, or any other part of the body that would need extra protein for healing purposes. Based on individuality, protein needs can use the range of 110-200% of the amount calculated for a healthy population.3

There are varying opinions about the amount of fat that should be included in the diet for the CF patient. Some sources recommended 20-30% calories from fat, while others recommend 35-40% calories from fat.2,3 If pancreatic enzymes are being taken with meals and the patient does not experience greasy stools or diarrhea, then absorption can become relatively close to normal and higher levels of fat may not be needed. This also depends on the growth of the individual patient, as well as fat stores and overall nutritional status. Among fat sources, the intake of omega-3 fatty acids (DHA and EPA) should be encouraged because they will help to reduce inflammation.1
For CF patients, carbohydrates are recommended to be within the same range as the rest of the population. An estimated 45-65% of calories from carbohydrates should be acceptable\(^1\). Along the USDA MyPlate nutrition guidelines for the entire population, half of carbohydrate sources should be whole grains, and carbohydrate sources high in fiber are recommended\(^2\). If CF-related diabetes (CFRD) develops, carbohydrate needs would remain the same, however the patient would have to count their carbohydrate servings at meals and would most likely need insulin therapy to manage blood glucose levels\(^1\).

**Micronutrients**

For CF patients, fat-soluble vitamins pose the largest concern out of all the micronutrients. Due to fat malabsorption issues, vitamins A, D, E, and K should be supplemented in the diet. These vitamins should be replaced with twice the normal RDA recommendation\(^1\). In order to ensure that the supplement will be absorbed, there are water-soluble brand sources of vitamins A, D, E, and K for this population\(^1\). The water-soluble supplement provides a higher concentration of beta-carotene in order to avoid the risk of a vitamin A toxicity. CF patients are still at risk for a vitamin D deficiency, and should be assessed annually on whether extra vitamin D is needed. Vitamin E will help prevent the neurological complications that can be seen in CF patients, and vitamin K needs should be met with the water-soluble multivitamin\(^2\).

Water-soluble vitamins, such as the B vitamins and vitamin C, can be met through the diet. Seeing a deficiency in water-soluble vitamins in CF is rare\(^2\). Taking a multivitamin every day should meet water-soluble vitamin needs\(^2\).

Minerals such as sodium, chloride, zinc, iron, calcium, magnesium, and fluoride may need to be supplemented. CF patients lose excessive amounts of sodium chloride through their sweat and need to supplement it. All infants with CF and an elevated sweat test need to
supplement 1/8 teaspoon (12.6 mEq) salt daily from birth to 6 months of age. After 6 months of age, 1/4 teaspoon (25.2 mEq) salt is needed for daily supplementation. More salt is recommended for those with CF that are physically active. Salt may be added to sports drinks (1/8 tsp to 12 ounces) to help replace sodium chloride losses\(^2\). Up to 4-6 grams of sodium chloride may be needed each day to replace sweat losses\(^1\).

Infants can lose excessive zinc in their stool with untreated pancreatic insufficiency, and may need 1 milligram of elemental zinc per kilogram of body weight per day for 6 months to replete, if symptoms such as severe diaper rash, lack of appetite, poor growth, dysgeusia, or compromised immunity are present. Zinc is also found in most multivitamins to meet daily needs if a deficiency is not suspected\(^2\).

Much of the general population can have their iron status affected by medications, blood loss, and dietary intake, however, when combined with malabsorption and inflammation in CF, patients can have iron deficiency anemia, anemia of chronic disease, or both. Iron supplementation may be needed\(^2\).

Calcium may need to be supplemented to meet daily recommendations for the entire population for adequate bone health and growth\(^2\). Magnesium blood levels should be monitored, and fluoride should be supplemented if it is not found in the local water\(^2\).

**Nutrition Support and Supplementation**

Nutrition support and dietary supplements are common among the cystic fibrosis population. Tube feedings are indicated if a child is experiencing weight loss or growth failure. The most common and convenient way for children to receive tube feedings is nocturnally, so they are not hooked up to a machine during the day and can freely attend school or activities. A nasogastric tube can be inserted and removed daily for short-term use\(^2\). The PEG tube is indicated for long-term tube feedings greater than a couple weeks, and is well tolerated in
patients who experience gastroenteritis reflux\textsuperscript{1}. With CF patients, parenteral (PN) nutrition is not recommended due to the higher risk of infection among this population\textsuperscript{1}.

Dietary supplementation is very common among children with CF due to increased energy requirements that cannot be met through diet alone. It is important to monitor any supplements that are not prescribed by the doctor, due to the possibility of adverse drug reactions and the unknown effects of supplements that have not been researched on the CF population\textsuperscript{1}.

**Special Diet Modifications**

Pancreatic enzymes need to be given before meals and supplements. For a mealtime longer than 30 minutes, the dose may need to be given before and halfway during the meal\textsuperscript{2}. For infants, enzymes should not be added to the formula in case all of the formula is not consumed\textsuperscript{1}. Among CF patients, lactose intolerance is common. Avoid milk during periods of intolerance with diarrhea and monitor to see if lactose intolerance continues\textsuperscript{1}. Monitor for intolerance of other foods and alter the diet accordingly\textsuperscript{1}.

Some research is being conducted regarding the benefits of adding turmeric and cumin to food for CF patients. Curcumin, a substance in turmeric, may directly stimulate CFTR chloride channels and have therapeutic effects\textsuperscript{1}.

Due to ever changing nutritional needs throughout various stages of life, it is recommended that cystic fibrosis patients have a complete nutrition assessment when they are diagnosed, and then yearly to follow-up with a Registered Dietitian\textsuperscript{2}. At every visit, CF patients will be screened to determine whether they are growing appropriately for weight and height. If the patient’s growth is not on track as predicted, the team will identify causes for the setback and help implement a plan\textsuperscript{2}. 
Presentation of the Patient

RW is a 9 year-old white male with no known allergies. He was born in September, 2004 with a history of failure to thrive (FTT) as an infant, and was diagnosed with cystic fibrosis in April, 2005. His CF diagnosis is the delta F508/R5553X genotype. The delta F508 mutation is a protein processing mutation, and it is the most common CFTR mutation that causes CF. With this mutation, the CFTR protein is made, but does not make it to the cell surface and is instead destroyed. Along with CF and FTT, he also has a past medical history of asthma, chronic rhinitis, pancreatic insufficiency, gastroesophageal reflux disease (GERD) with Nissen fundoplication and several repairs due to reflux, malnutrition, adenotonsillectomy, tympanostomy tubes, and a previous gastrostomy tube placement with removal in April, 2013.

During an outpatient visit on January 23, 2014, the disciplinary team saw the patient. At that time, his anthropometric measurements were taken. His weight was 25.31 kilograms at the 15th percentile, height was 128 centimeters at the 12th percentile, and BMI was 15.5 at the 31st percentile. RW had gained 1.21 kilograms over the past month since his last outpatient visit. He had personally requested to replace a gastrostomy tube back in for supplementation. He does not feel like a normal kid due to a large number of supplemental liquids during the day and feels like an outcast from peers and friends. He would prefer to have nocturnal feedings so that he does not need to consume so many liquid calories during the school day. After meeting with the patient and a parent, the disciplinary team supports the placement of a gastrostomy tube.

His recent stay in the hospital was from January 29 to February 12, 2014. On this hospital stay, he came into the hospital with shortness of breath, cough, wheezing, and declining weight due to non-compliance with taking nutritional supplements and medications. On admission, his anthropometric measurements were taken. His weight was 24.9 kilograms between the 10th-25th percentiles, height was measured to be 128 centimeters between the 10th-
25\textsuperscript{th} percentiles, and BMI was 15.2 at the 25\textsuperscript{th} percentile. His ideal body weight (IBW) for his age of 9.25 years old is 26.7 kilograms and he is 93\% of his IBW for his height. From the recent outpatient visit, the interdisciplinary team had also planned to replace his feeding tube, which will be surgically placed during this hospitalization.

As part of the interdisciplinary team, the Registered Dietitian (RD) had ordered the patient supplemental carnation instant breakfast to be mixed with chocolate ice cream three times per day, due to poor intake and reported declining weight. On my visit with the patient, he agreed that he likes his chocolate milkshakes, but he does not like yogurt and only ate a couple bites of his cinnamon roll for breakfast.

Family history from this patient is somewhat extensive and complex. RW has a 1-year-old half-sister, 3-year-old half-brother with mild CF, 4-year-old half-sister, 8-year-old sister, and 13-year-old half-brother. His biological father lives in Arizona, and his mother is currently going through a divorce with her second husband after verbal and physical altercations. Through the divorce process at the current time, the mother’s female partner has moved into the home, as well as the husband’s girlfriend. As all of these issues are occurring at home, the patient has been expressing possible symptoms of depression and anxiety. Lately the patient has been frustrated and angry towards his mother, and his grades went down during the second semester at school. He has witnessed some of the recent altercations in the home. He also has been stating “I wish I was a normal kid” and according to the patient, at times he wishes he would not wake up. RW’s family issues intertwined with feeling out-casted from friends for being “different” is likely another source of poor food intake recently.

Although RW has a complicated life at home, he does go to school when he is able and is followed by his school counselor. The patient is in third grade and enjoys playing floor hockey. He also enjoys eating funnel cake and playing video games.
Discussion of Medical/Surgical Hospital Course

RW’s hospital stay was from January 29 to February 12, 2014. During this hospital stay, he presented with shortness of breath, cough, wheezing, and declining weight due to non-compliance with taking nutritional supplements and medications. From a recent outpatient visit, the interdisciplinary team had also planned to replace his feeding tube next week.

The RD had seen the patient shortly after admission to offer snacks and supplements to increase food intake. The RD ordered Carnation instant breakfast to be mixed with chocolate ice cream and served to patient three times each day. On February 4, the patient confirmed that he likes his Carnation instant breakfast milkshakes, but is not eating much of the food he has been receiving.

On February 10, the patient received a percutaneous endoscopic gastrostomy (PEG) tube due to the past several weeks to months of having issues with inadequate food intake and questionable depressive symptoms, which could be an added reason for suboptimal food intake. There were no complications with the procedure.

On February 11, lab results showed many normal results, with some slightly abnormal lab values also present:

- Sodium (Na) 140, Potassium (K) 4.1, Chloride (Cl) 107, Calcium (Ca) 9.1, Carbon Dioxide (CO₂) 27, Anion gap 6, Creatinine 0.44, Blood Urea Nitrogen (BUN) 16, Blood Glucose (BG) 121↑, Albumin 4.2, Total protein 6.8, Aspartate Aminotransferase (AST) 26, Alanine Aminotransferase (ALT) 120↑, Total alkaline phosphatase 187, and Gamma-glutamyl transpeptidase (GGT) 78.

Other tests performed on the patient included:

- Complete Blood Count (CBC): results were within normal limits
- Basic Metabolic Panel (BMP): results were within normal limits
• Chest X-ray: Showed persistent asymmetric bibasilar interstitial infiltrate consistent with atelectasis and/or pneumonia (PNA) with consolidation, now better visualized in the right posterior costophrenic angle on the lateral view consistent with PNA. Underlying viral bronchiolitis and/or reactive airway disease exists.

The patient was discharged on February 12, 2014. On discharge, diagnoses included CF exacerbation, pneumonia, nutritional failure, and adjustment disorder. Due to the more frequent episodes of expressing depression and anxiety symptoms, the medication Ativan was added to his medication list, to be used every 12 hours as needed for severe agitation. The doctor would also like for RW to try counseling and see if this helps him overcome some emotional struggles at the current time. The doctor will order an antidepressant, Prozac, if the patient does not improve with counseling.

Also after discharge, the patient will begin to receive his nocturnal tube-feedings to supply the majority of his estimated nutrition needs. Elecare Junior will be used, since it is an elemental formula and PERT enzymes do not need to be taken with this type of formula. Water flushes of 30 milliliters are recommended before and after each tube feeding. PERT enzymes will need to be continued with meals or snacks during the day. RW’s AquADEK vitamins and vitamin D supplement will be continued on discharge as well.

On returning home, RW is encouraged to keep adding salt (about ¼ teaspoon) to his meals and to keep eating salty foods to replace sodium chloride. Supplements such as Boost Kids Essentials 1.5 in chocolate (360 calories), 1.5 ounce Benecalorie in meals (330 calories), and Carnation instant breakfast mixed with ice cream or whole milk were suggested to the family as options for RW to keep up his caloric intake during the day. RW will follow-up two weeks after discharge with his Pediatric gastroenterologist.
Discussion of Nutrition Care

There are many ways to assess the nutritional needs of a pediatric cystic fibrosis patient. For RW, multiple equations were calculated for comparison, to come up with the most appropriate range of caloric needs. First, the World Health Organization (WHO) equation was used for a male age 3-10 years old \([22.7W + 495]\), where \(W\) is weight in kilograms, and the result was multiplied by an adjustment factor of 1.7 for an active child requiring either catch-up growth or severe stress\(^7\). This result from the WHO equation was also multiplied by 1.2 in order to account for 120\% of caloric needs\(^1\). The final result was 2,162 calories. The Schofield equation was also used for a male age 3-10 years old \([(12.6W) + (1.303H) + 414.9]\), where \(W\) is weight in kilograms and \(H\) is height in centimeters\(^7\). The result was multiplied by an adjustment factor of 1.7, and then 120\% of this result was taken\(^1\). The final result for the Schofield equation was 2,181 calories. In order to estimate a range of nutrition needs, 150\% of caloric needs were also calculated for each equation. With all factors considered, RW’s estimated caloric needs are 2,150 to 2,700 calories, based on 120-150\% of the WHO and Schofield equations\(^1,7\).

Protein needs were calculated based on 1.5-2.0 grams of protein per kilogram of body weight per day. Weighing 24.9 kilograms, RW needs a minimum of 38 to 50 grams of protein per day\(^7\). However, with a diet consisting of 2,150 calories, consuming 20\% of calories from protein would provide RW with 107 grams of protein per day\(^2\). Since RW will likely experience some malabsorption due to his disease, does not have kidney issues, and is 93\% of his ideal body weight, then this amount of protein is entirely acceptable for him, as long as he is meeting his minimum recommendation.

Fluid needs are calculated based on the Holiday-Segar Method for pediatric patients. CF patients have no excess needs for fluids compared to the healthy pediatric population. The
Holiday-Segar Method equation for children over 20 kilograms is $1,500 \text{cc} + 20 \text{cc/kg} \times \text{kg} > 20\text{kg}$.

Based on this equation, RW needs 1,600 milliliters of fluids each day, or about 7 cups. More fluid may be needed for repletion in active children playing sports.

The doctor prescribed the regular pediatric diet for ages 4-14 for this patient. Supplements were also ordered, which included Carnation instant breakfast to be mixed with chocolate ice cream 3 times per day. The doctor prescribed Zenpep pancreatic enzymes to be taken at meals (3) and with supplements (2). RW will continue eating salty foods and adding salt to his meals (about $\frac{1}{4}$ teaspoon) to replace sodium chloride losses. The patient is also ordered to take a vitamin D supplement (5,000 IU per day) and a water-soluble multivitamin containing vitamins A, D, E, and K (AquADEK vitamins). Two softgels of AquADEK vitamins provide RW with 36,334 IU of vitamin A (92% beta carotene), 300 IU of vitamin E, 1,600 IU of vitamin D, 1,400 micrograms of vitamin K, and 20 milligrams of zinc per day.

Nutrition education was not appropriate at this time, as no parents were present when I visited with the patient. However, the Registered Dietitian and myself did encourage RW to eat the food coming on his trays, attempted to get some food preferences from him, and also encouraged him to drink his supplements. The patient does like his chocolate “milkshake” supplements and according to the nursing staff he seems to be drinking all three each day. With a PEG tube being replaced, the patient will go home on nocturnal tube feeding. Elecare Junior is the elemental formula that will be used for RW, since PERT enzymes do not need to be taken with this type of formula. The tube feeding will run for 12 hours from 7pm to 7am via a continuous pump at 100 milliliters per hour to provide 1,200 milliliters of formula, and a 30 milliliter flush before and after each tube feeding. The formula will be concentrated to 45 calories per ounce of formula by using 915 milliliters (31 ounces) of water with 41 scoops of formula powder to provide 1,833 calories and 56 grams of protein each feeding.
RW will still need to take his enzymes during the day with meals or snacks. He can consume any supplements he had previously been drinking, such as the 8-ounce Boost Kids Essentials 1.5 in chocolate (360 calories each), Carnation instant breakfast milkshakes, or add 1.5-ounce Benecolorie to meals (330 calories each) if he is not eating enough during the day. Since the nocturnal tube feeding will be meeting most of his needs, the supplements may not be necessary at this time. The patient will continue to follow-up with the Pediatric Registered Dietitian at his outpatient appointment visits.
Summary

Since RW had the percutaneous endoscopic gastrostomy tube replaced, our goal is to see him tolerate the tube feeding and reach 100% of his ideal body weight to optimize pulmonary function. Although it may seem extreme to place a feeding tube in a young child, it can be more convenient for some people and may increase the quality of life for the patient. For RW, he will be receiving the majority of calories overnight, so he can hopefully feel like a “normal” kid at school and drink chocolate milk, instead of chocolate supplements, during lunchtime.

In conclusion, cystic fibrosis is a chronic disease that affects primarily the lungs and pancreas, as well as the GI tract, bile ducts and liver, exocrine glands, and many other body functions. An interdisciplinary team is needed to help manage this disease and the complicated needs of each patient. An early diagnosis is key for better outcomes and allows for nutritional management to begin immediately so malabsorption can be recognized and malnutrition can try to be avoided. The dietitian plays a large role in the team to make sure the patient receives optimal nutrition at all times to maximize growth from infancy to childhood, to adolescence and adulthood, and to optimize pulmonary function. Thankfully, with effective pancreatic enzyme replacement therapy, nutritional supplements, and water-soluble vitamins and minerals, it is becoming much easier to manage the nutritional needs of cystic fibrosis patients. Hopefully the predicted median age for these patients will reach past the early forties, as other medications and treatments emerge in the years to come.
Medication Bibliography

At the time of discharge on February 12, the medication list included many pulmonary medications, a laxative, enzymes, vitamins with minerals, and more. Many of the pulmonary medications did not have any nutrition implications or food-drug interactions.

RW is prescribed Zenpep, also known as pancreatic enzymes, pancrelipase, or pancreatin. The enzymes are a combination of porcine lipase, amylase, and protease to help with digestion and absorption of nutrients in the intestines. The medication is used to treat steatorrhea due to pancreatic insufficiency. Dosage is based on symptoms and should be taken with food with a pH less than 4.5. RW is prescribed Zenpep 20, providing 20,000 lipase units per capsule. He is prescribed to take 3 capsules per meal, providing 60,000 units of lipase per meal, which is 2,370 units of lipase per kilogram of body weight per meal. Taking these enzymes can decrease iron and folate absorption.

Polyethylene glycol, commonly known as MiraLax, is taken twice per day. Polyethylene glycol is an osmotic laxative that can cause bloating, cramps, flatulence, and diarrhea. A high fiber diet with plenty of fluids is recommended to help prevent constipation. Pantoprazole, also known as Protonix, is taken once per day. Pantoprazole is proton-pump inhibitor, used as an anti-GERD and anti-secretory drug to help with reflux symptoms. Taking this drug may decrease absorption of vitamin B\textsubscript{12} and iron.

RW is taking loratadine, also known as Claritin, once per day. Loratadine is an antihistimine and can increase appetite, increase weight, increase thirst, and can also cause anorexia symptoms. RW also takes montelukast, also known as Singulair, once per day. Montelukast is used for asthma prophylaxis and as a chronic treatment. Montelukast has no interactions with grapefruit or citrus fruits, however it can cause some indigestion and diarrhea. Every 4 hours, RW must take 2 puffs of albuterol, also known as Proventil. He must also take 2
inhalations twice per day with his vest therapy. Albuterol is an anti-asthma bronchodilator and can cause increased appetite or anorexia-like symptoms\(^8\).

Due to recent depressive symptoms, the doctor may be adding Ativan to the medication list, only to be used as needed for severe agitation. Ativan is a benzodiazepine drug used for antianxiety and antipanic. While taking Ativan, caffeine should be limited and grapefruit and related citrus fruits should be cautioned. Ativan can cause decreased weight, anorexia, and increased thirst. In the event that RW does not respond to counseling for depression, the doctor will order Prozac, also known as fluoxetine. Prozac can also produce anorexia-like symptoms with a decreased weight.

RW will also continue with his over-the-counter vitamins. Each day he is taking 5,000 IU of vitamin D, also known as Ergocalciferol. Vitamin D causes increased calcium absorption, and can produce anorexia-like symptoms, decreased weight, and increased thirst. Vitamin D levels should be monitored for deficiency or possible toxicity\(^8\). He is also taking 2 soft-gels of AquADEK vitamins each day to provide 36,334 IU of vitamin A (92% beta carotene), 300 IU of vitamin E, 1,600 IU of vitamin D, 1,400 micrograms of vitamin K, and 20 milligrams of zinc per day\(^2\).
References


