Cystic Fibrosis: Living With a Chronic Disease

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Cystic Fibrosis: Living With a Chronic Disease

Brittany Heidenreich

Carroll College
SIGNATURE PAGE

This thesis for honors recognition has been approved for the Department of Nursing.

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Abstract

Cystic Fibrosis (CF) is an inherited chronic disease that shortens the life expectancy of the individual to an average age of 37. It is a debilitating disease that negatively affects the respiratory system and digestive system. The purpose of this thesis is to explore the lives of persons living with CF. Two males and two females with CF were interviewed as well as a parent who has a child with CF. A total of five individuals volunteered to be interviewed coming from a variety of different states including Washington, Montana, Idaho, Illinois, and Oregon. Phenomenology method was in order to explore the lives of individuals living with CF. Closed-session interviews were conducted to gain insight into living with CF. The major themes were utilizing family support as a coping strategy, time-consuming treatments, decreasing energy, and staying positive. Participants expressed relying highly on family members to help them cope with their disease and were open with their families when discussing concerns about their disease. Declining energy and health along with time-consuming treatments were common discouraging aspects of CF that required having a positive attitude and utilizing support systems to continue treatments. These findings cannot be generalized to all individuals with cystic fibrosis.
Acknowledgements

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Dedication

This thesis is dedicated to my niece, Brooklyn, and the senior nursing class of 2009.
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Chapter I

Living With a Chronic Disease

Cystic Fibrosis is most common among Caucasian people and is fatal with no cure (Berge, Patterson, Goetz, & Milla, 2007). It affects the respiratory and digestive systems starting at birth and continues to affect the individual throughout life. Cystic Fibrosis (CF) can cause “life-threatening lung infections and obstructs the pancreas and stops natural enzymes from helping the body break down and absorb food” (Cystic Fibrosis Foundation [CFF], 2007c). Infants who are diagnosed with this disease have a difficult time breathing and gaining weight starting from day one of life. CF is an inherited chronic disease that shortens the life expectancy of all who are diagnosed with this debilitating disease. According to the CF Foundation, in the 1950s children were not expected to live to reach elementary school, but in 2006 the average life expectancy for people with CF was reported to be more than 37 years old. Currently there are about 30,000 Americans, children and adults, who are diagnosed with CF. Every year approximately 1,000 new CF diagnoses are made, and 70% of these individuals are diagnosed by age two (CFF, 2007c).

Through research, nurses can develop a better understanding of this disease and will therefore be able to empathize and help those who are affected. It is important that nurses as well as other health care providers understand CF and the impact that this disease has on the lives of individuals affected by it. The purpose of this thesis is to explore the lives of persons with CF.
Significance

Although the life expectancy has increased dramatically in the last 50 years, it is still far below the average life expectancy of people without this disease. It is important for nurses to become familiar with this disease because they can play a very important role in treatments and support because “the client with CF needs daily therapy to slow disease progress and enhance gas exchange. There is no cure for CF” (Ignatavicius & Workman 2006, p. 608). The nurse’s role is to help with therapy and to teach the individual as well as his or her family how to implement the therapies so that they are able to be effective in homecare (Taylor, Lillis, & LeMone, 2005).

Impacts of CF on the Individual

Since CF has no cure, treatment plans are individually made to slow the progression of the disease. This has a huge impact on an individual with CF over time. Some treatments include breathing techniques and self-clearing of the lungs, done by clapping the chest; this is called chest physiotherapy. Although treatments are necessary and very helpful, “this management often becomes onerous over time with many young adults finding it increasingly difficult to fulfill their aspirations for a ‘normal’ life” (Badlan, 2006, p. 264). For individuals with CF, their treatment plans are no longer just treatments; instead they become a way of life. Since recent technology has extended the life expectancy age to 37, individuals in their 20s and 30s with CF are not familiar with the idea that they have extra years to live because they were told that their life expectancy was much lower (Badlan, 2006). One individual, in a study done by Badlan, expressed that there was difficulty in living a normal life because with CF an individual has “two states ‘when you are well and when you are not well’ ” (2006, p. 267). As a result of life-
long treatments, individuals with CF have a difficult time living a “normal” life; this also may lead to psychosocial issues such as low self-esteem, impaired physical appearance, and depression related to being different from other children (Badlan, 2006).

Following treatment plans throughout a lifetime can be difficult physically and emotionally to an individual with CF. Another factor influences the ability to follow treatment plans, and that is financial costs for treatments and medications. Costs can be overwhelming to parents who have a child with CF, but there are programs designed to ease the stress of financial costs. Medicaid, State Children’s Health Insurance Program, the Food Stamps Program, and Supplemental Food for Woman, Infants, and Children (WIC) are all resources that are available to families who have a child with CF. There are also assistance programs for adults and elderly who have CF. These include Medicare, Social Security, and many others (CFF, 2007a). Clinics in the United States have reported that children who belong to a family with no health insurance or low socioeconomic status are associated with poorer outcomes with medications and less success slowing the disease process (Schechter, Shelton, Margolis, & Fitzsimmons, 2001).

Families who utilize Medicaid are of a lower socioeconomic status (SES) than those who utilize private insurance. A study done by Schechter and Shelton 2001) compared families who are of low socioeconomic status (those who used Medicaid) to those who use private insurance. The study concluded that “medically indigent CF patients suffer more serious consequences of their disease than does the general CF population. They have more than a threefold greater risk of death, and survivors have significantly worse pulmonary function and growth”
(Schechter & Shelton 2001, p. 1331). Although the study compared those who use Medicaid to those who have private insurance, the study also concluded that other factors signify having a low socioeconomic status other than utilizing Medicaid. One factor that is associated with low socioeconomic status (SES) is lack of adequate nutrition, which worsens pulmonary function and contributes to an even lower weight class than other children with CF who do not have a low SES. Another important factor associated with low SES is smoking. Low SES families are more likely to have smokers in the family. Cigarette smoke decreases pulmonary function and increases the risk of contracting a respiratory virus (Schechter & Shelton, 2001). CF is expensive to treat and even with programs such as Medicaid, families may not be able to afford the best treatments and medications available.

Lung transplantation and/or pancreatic transplantation are a last resort treatment for individuals with CF. These procedures are extremely beneficial and may extend life by 10 to 20 years, but transplantations do not cure CF because CF is caused by a genetic defect that has no reversal treatment. Lungs are scarce and surgical procedures can be very dangerous for individuals with CF because the disease process causes problems in many organs in the body (Ignatavicius, 2006). With all surgeries, there are benefits and risks, so it is important to determine if surgery is right for a specific individual with CF.

Summary

A thousand children are diagnosed with CF every year, and many will not live to have children or watch their children grow up. This is a result of both the disease process and cost of medications and treatments. Treating a genetic disease for a lifetime is expensive and difficult to keep up with. The purpose of this thesis is to explore the lives
of individuals with CF and become more aware of the impact of the disease on the lives of individuals and families affected by CF. There is no cure for CF, so treatment plans are crucial to slowing the disease process. Nurses play a role in teaching treatment plans and helping families implement them once they are established; therefore it is vital that nurses have an in-depth understanding of what it means to live with CF.
Chapter II

Review of Research

CF is an inherited disease that primarily affects the respiratory and digestive systems in the individual’s body. There is specific pathology for the respiratory and digestive systems and how they are affected with CF. This section will critique the research on etiology, pathology, diagnostics, treatments, and the lived experience of CF.

Etiology and Incidence

Cystic fibrosis is an inherited disease that affects many organs in the body which makes the disease more complicated and difficult to manage. The CF gene was identified in 1989; as a result the 1990s became a break-through decade in understanding CF and the manifestations that present. A single mutation on chromosome 7 was identified that “encodes for the cystic fibrosis transmembrane regulator (CFTR), which functions as a chloride (Cl⁻) channel in epithelial cell membranes” (Porth, 2007, p. 506). A mutated CFTR gene is only found in individuals with CF and causes many unwanted manifestations in the individual with CF (Gibson, Burns, & Ramsey, 2002).

It is impossible to predict the first time CF will occur within a family because carriers of the disease do not present with any signs and symptoms. CF can only be expressed if both parents are carriers. If both of the parents are carriers, this does not mean that their child will automatically inherit CF. Since the gene is recessive, the fetus only has a one in four chance of inheriting CF. In most cases the parents do not know that they are carriers of the mutated gene; therefore there is little possibility of prevention with this inherited disorder (CFF, 2008). Cystic fibrosis can manifest in anyone who receives the two recessive genes from his or her parents. Race is a factor in determining
the incidence rate of CF. This disease presents most often in Caucasians and is rare in Asian and African American populations. Age is not a factor because CF is inherited and affects the person from birth throughout the lifespan. Gender is also not a risk factor because CF affects males and females equally (Ignatavicius & Workman, 2006). Although CF is not curable, it is treatable; therefore it is important to understand what happens to the individual’s body with CF so that it can be managed appropriately.

**Pathology**

Symptoms of CF manifest in many areas of the body making the disease difficult to live with. Both the respiratory and digestive systems are affected greatly in individuals who have CF. In addition to respiratory and digestive manifestations, the individual has an increased amount of sodium in the body and on the epithelium. Pathology of the respiratory system, digestive system, and sodium production will be explored throughout the pathology sections. CF can cause the individual to develop other diseases such as diabetes mellitus, a result from reduced pancreatic function, as well as serious respiratory infections (Boucher, 2006).

**Respiratory.** According to a study done by Boucher, respiratory infections in children with CF are resistant to antimicrobial therapy; therefore the infections are very difficult to eradicate and may not ever be completely eradicated. As a result of a mutated CFTR gene, reabsorption of sodium (Na+) and chloride (Cl⁻) does not occur, causing high amounts of NaCl. There are many hypotheses on how this increase in NaCl affects the respiratory system. When the CFTR gene does not work correctly, Na+ is absorbed with no restraint and Cl⁻ is not secreted properly, causing an increase in absorption of liquid in the airways so that the airways become dehydrated. This dehydration of the
airways allows mucous to attach to the airways’ surfaces causing airway obstructions and chronic infection (Boucher, 2006). Respiratory problems are progressive, and as a result they become more frequent and cause acute and chronic exacerbations causing the individual to “have chest congestion, limited exercise tolerance, cough, sputum production, use of accessory muscles, and decreased pulmonary function” (Ignatavicius & Workman, 2006, p. 608).

Digestive. The digestive system is also affected in individuals with CF; as a result the adult client with CF is usually smaller and thinner than average. The pancreas is affected by the mutation of the CFTR protein causing the pancreas not to function correctly. According to Luder (2003), “exocrine pancreatic insufficiency is present in 85% to 95% of CF patients” (p. 93). Pancreatic insufficiency is caused by an obstruction of the ducts leading to the pancreas. An obstruction is caused by an increase in highly concentrated secretions from the pancreas, which eventually causes fibrotic tissue. An obstruction in the ducts causes a decrease in digestive enzymes being released and decreased digestion (Luder, 2003).

Other manifestations of the digestive system include abdominal distention, gastroesophageal reflux, rectal prolapse, foul-smelling stools, and steatorrhea. The client may also be malnourished and have many vitamin deficiencies” (Ignatavicius & Workman, 2006, p. 608). In later stages of CF, a loss of body mass can occur which correlates with impaired pulmonary function.

Body Composition. A study done by Ionescu, Chatham, Davies, Nixon, Enright, and Shale found that “loss of skeletal muscle mass would include inspiratory muscles and impair their function” (Ionescu et al., 1998, p. 1271). The researchers also found “a clear
relationship between body composition and impairment of inspiratory muscle function in adults with CF” (Ionescu et al., 1998, p. 1274). This study has limitations, but does still “support the hypothesis of a link between inspiratory muscles function and body composition in CF” (Ionescu et al., 1998, p. 1275). To test this hypothesis they “determined the effect of body mass index (BMI) and lean body mass index (LBM) depletion on handgrip (HG) force and inspiratory muscle function (IMF)” (Ionescu et al., 1998, p. 1271). Forty-nine patients were used to find a correlation between IMF and BMI and 25 patients were used to find a correlation between IMF and LBM. A person with low LBM would be likely to have impaired pulmonary function of the inspiratory muscles as well as reduced handgrip. The study also concluded that these persons would also have a reduction of skeletal muscle function in general, not just reduction in inspiratory muscles. This study does have limitations which include the difficulty to compare pulmonary function of one individual with CF to another individual with CF, because of the differences in manifestations of the disease as well as the person’s age. Understanding the link between one complication of CF with another complication can be crucial for nurses to implement interventions. People will have complications such as decreased BMI and decreased food absorption that increases the need for nurses to understand all manifestations of CF.

Sweat production. An excessive amount of sweat is produced in individuals with CF as well as a defect in chloride absorption which allows excessive sodium to be released through sweating (Ignatavicius & Workman, 2006). As a result of this excessive production, CF can be diagnosed by using a sweat test. The sweat production is caused by a mutation in the CFTR. Production of sodium is not affected in individuals with CF;
instead reabsorption of sodium (Na+) and chloride (Cl⁻) does not occur causing an increase of NaCl in the sweat in individuals with CF (Porth, 2002).

*Diagnostic testing.* Early detection of CF is crucial. The earlier CF is detected, symptoms such as malnutrition can be controlled. A small number “around 10% are diagnosed at birth because of meconium ileus, but the majority are diagnosed in early infancy because of recurrent lower respiratory tract infections or malnutrition or both” (Koch & Hoiby, 2000, p. 239). The test used most frequently, considered the gold standard test of CF, is the sweat test. The sweat test is ordered when the individual shows signs and symptoms of CF. An excess of sweat indicates CF. The Cystic Fibrosis Foundation (CFF) provides accurate and adequate information on all aspects of CF including the sweat test. According to CFF, the sweat test is painless and does not require the use of needles. A normal level of less than 40mmol/L indicates no diagnosis of CF. A range from 40-60 mmol/L indicates that the individual is borderline and may require a retest at a later date. A diagnosis of CF is given only when the results are 60mmol/L or above (CFF, 2007b).

In 1989 the CF transmembrane regulator gene (CFTR) was identified. In order to detect this CFTR gene, the mutation ΔF508 is tested for. The problem with this is that about 70% of CF mutations worldwide are ΔF508, leaving about 30% a different mutation type. There are some cases in which the individual is misdiagnosed; therefore the individual being tested should be tested twice on different occasions. The test can also be more accurate by “testing on both arms simultaneously and with inclusion of quantification of potassium – besides chloride and/or sodium” (Koch & Hoiby, 2000, p. 239). The test is used worldwide and when done correctly is very accurate.
Treatments

CF is a disease that requires many medications and complementary therapies to slow progression because it affects multiple organs in the body. Pharmacological treatments will be used frequently and are specific to system as well as level of exacerbation. Specific medications are used in acute exacerbations versus maintenance therapy drugs. Complementary therapies are also available to improve treatment of CF which includes diet and chest physiotherapy. A surgery option, lung transplantation, will also be addressed in this section. An analysis of treatments will be in this section in order to provide an insight into whether treatments are working and whom they are effective for (CFF, 2005).

Secretions. The lungs can be affected very severely in individuals with CF inhibiting the function and creating an excess of mucous production. Dornase alfa, also called Pulmozyme, is used to help break up the thick mucous secretions produced in CF. Dornase alfa is inhaled by mouth using a nebulizer that is prescribed by a primary care provider. Breaking up the thick secretions will increase the airflow and decrease the risk for infection buildup (MedlinePlus, 2008).

Pulmonary infection. Serious infections can develop in the lungs starting from bacteria such as *Staphylococcus aureus*, *Haemophilus influenza*, and *Pseudomonas aeruginosa*. For mild exacerbations of *Staphylococcus aureus* infections, Azithromycin, an antibiotic, is used (Micromedex, 2008). For mild exacerbations of *Haemophilus influenzae* infections, Amoxicillin, Cefaclor and Azithromycin antibiotics are used (Micromedex, 2008). For more serious exacerbations, stronger antibiotics may need to be used. Severe exacerbations of *Staphylococcus aureus* are treated with many antibiotics
including Cefazolin Sodium, Naficillin Sodium, and Vancomycin Hydrochloride (Micromedex, 2008). Cefazolin Sodium and Naficillin Sodium are used to treat severe infections such as those of blood, heart valve, bone, skin, and joint and must be given through an injection or with intravenous fluid. Vancomycin Hydrochloride, an antibiotic, must be added to intravenous fluid in order to be effective (MedlinePlus, 2008).

*Digestive system medications.* Although respiratory impairment can be the most severe symptoms of CF, the digestive system is also affected greatly. The pancreas is specifically impaired with CF and this causes difficulties because “the pancreas produces enzymes that are required for the digestion and absorption of food” (Micromedix, 2008).

In order to increase effectiveness of the digestive system, individuals with CF must take supplemental pancreatic enzymes. Similar to the respiratory system “a thick, sticky mucus forms in the pancreas. The mucus clogs the pancreatic duct and prevents the enzymes from reaching the small intestines” (Cystic Fibrosis Services Pharmacy Educational Series, 2007, p. 1). Related to the lack of enzymes, individuals with CF are at a large risk for mal-absorption which can lead to malnutrition and inability to maintain a stable weight. With an increased risk for malnutrition there is also an increased risk for lung disease because “nutrition and lung health are closely linked. Good nutrition helps maintain breathing muscle strength” (Cystic Fibrosis Services Pharmacy Educational Series, 2007, p. 1). The enzymes taken are specifically made to dissolve in the small intestine. They are enteric coated so that they are not digested in the stomach when taken by mouth. Pancrecarb, Creon, Minimicrospheres, Ultrase, and Pancrease are all recommended brand-name pancreatic enzymes for individuals with CF (Cystic Fibrosis Services Pharmacy Education Series, 2007, p. 2).
Education is crucial for diagnosed individuals with CF because without education the disease cannot be treated to full potential. Basketter, Sharples, and Bilton (2000) analyzed adults’ knowledge of supplementation to determine if they were correctly administering pancreatic enzymes. Sixty-five adult CF patients 15-50 years old were sent a questionnaire. The questionnaire asked them to record details of their own supplemental use. It assessed these areas: “the type of enzyme preparation, total daily intake, number and timing of enzyme with average meals and snacks and number they would take with specified meals” (Basketter, Sharples, & Bilton, 2000, p. 353). Fifty questionnaires were returned. The results concluded that individuals were not adjusting amount of enzymes needed to varying fat content as they should be. Through this study, nurses and other healthcare workers can determine that there is a need to increase education regarding enzyme intake to the adult population 15-50 years of age (Basketter, Sharples, & Bilton, 2000).

Complementary Therapies

Complementary therapies are used in addition with medications and/or surgery in order to allow healthcare providers to deliver optimum treatment to all who are ill. Dietary measures are taken to ensure that those with CF are consuming a diet high in calories. Chest physiotherapy has now become a standard of treatment for individuals with cystic fibrosis. With the addition of complementary therapies to pharmaceuticals and surgeries, healthcare providers and individuals with CF are able to take several approaches to treatment.

Dietary measures. In addition to pancreatic enzymes, an individual with CF can also maintain body weight by eating a high fat diet. This may be difficult for individuals
with CF because of a lack of appetite; it may be easier to eat many small meals a day instead of large meals. It is important to get the needed caloric intake because CF causes individuals to have less energy, so eating foods "from all of the food groups, which includes fruits, vegetables, breads, dairy products, meat and fish" (Micromedex, 2008, p. 1) may help increase energy (CFF, 2007).

_Chest physiotherapy_. Several techniques including Postural Drainage (PD), Percussion (P), and vibration are used in addition with medications to break up the thick, sticky mucus that builds up in individuals with CF. Breaking up this mucus not only increases the effectiveness of breathing but it also reduces the incidence of infection. Patients should use medications in concurrence with P and PD to increase the effectiveness of chest physiotherapy; "bronchodilators should be taken before PD & P to open the airways. And aerosolized antibiotics should be taken after PD & P to treat the opened airways" (CFF, 2005, p. 1). The right lung is composed of three lobes, and the left lung is composed of two lobes and "each segment of the lung contains a network of air tubes, air sacs and blood vessels" (CFF, 2005, p. 1). It is essential for exchange of oxygen to occur between the blood and air.

The goal of PD is to use gravity to get the mucous moving out of the lungs. The individual sits and lies in different positions such as Trendelinburg, where the head of the bed is lowered below the rest of the bed, to utilize to force of gravity to help clear the lungs. The nurse or the performer should slowly lower the head of the bed "so that the client’s head is positioned at no greater than 25° downward angle" (Smith, Duell, & Martin, 2008, p. 955).
Percussion is done after PD, by cupping the hand and clapping with the palm face down to the individual’s chest, back, and sides around the lungs. Each area should be percussed for 3—5 minutes (Smith, Duell, & Martin, 2008). Percussion can be done on children by respiratory therapy, a parent, a sibling, or by anyone else. When individuals reach an adolescent or adult age, they can be taught to do it to themselves. In order for percussion to be effective, it must be done “vigorously and rhythmically, but should not be painful or sting if the hand is cupped properly” (CFF, 2005, p. 2). If done correctly, percussion should make a hollow sound. Areas such as the spine and breastbone should be avoided and percussion should not be done over bare skin because it may cause pain. The goal of PD and P is to move the thick mucus to the larger airways so that the mucus can be coughed out. Without moving the mucus to the larger airways, it becomes stuck in the small airways and cannot be removed by coughing (CFF, 2005). After percussion is complete, it should be followed by vibration, coughing, and deep breathing (CFF, 2005).

Vibration is performed by keeping the arms straight and the wrists stiff so that the vibrations can be delivered in bursts. The individual receiving the vibration must be instructed to inhale deeply and then exhale using pursed lips. During the exhale, the performer will then begin contracting and relaxing the arms at a fast pace several times. Vibration should be done for 3 to 4 exhales over each area (Smith, Duell & Martin, 2008). Overall, chest physiotherapy is very helpful for individuals with CF because the thick, sticky mucous can be broken apart and removed by coughing more easily.

_Surgical Treatment_

Lung transplantation and/or pancreatic transplantation is a last resort treatment for individuals with CF. The procedures are extremely beneficial and may extend life by 10
to 20 years. Transplantations do not cure CF because CF is caused by a genetic defect that has no reversal treatment. With all surgeries there are risks and benefits. Lung transplantation is very expensive, and healthy lungs, used for transplantation, are scarce.

_Lung transplantation._ A questionnaire-based pilot study written by, Yorke, Parle, James, Gay, Harkness, & Glanville (2006) was used to compare pre-transplant and post-transplant groups to determine the difference in quality of life for each group. The individuals who had already received a transplant (the post-transplant group) scored higher in physical function, physical role, bodily pain, general health, vitality, social functioning, role-emotional, and mental health than the pre-transplant group. The setting chosen was a major lung transplant unit in Australia. Twenty-seven individuals with CF were chosen for this study and split into two groups, nine for pre-transplantation and 18 after transplantation. According to the results of this study, it is apparent that the nurse’s role is to identify any extra needs that people awaiting a transplant might need. It is also important for nurses to direct interventions towards these needs in order to provide the highest quality of comfort and care. The categories of anxiety, depression, and total distress did not show significant differences between pre- and post-transplant individuals. The study was conducted because little is known about the transition period for young adults awaiting transplantation (Yorke, Parle, James, Gay, Harkness, & Glanville, 2006).

**Analysis of Treatment**

Several studies, such as the Quality of Life measurement and the Survivorship model, were conducted to determine how well treatments are working for individuals with CF. The Quality of Life measurement measures whether treatments improved the quality of life of the individual with CF. The Survivorship model was conducted to
analyze if treatments lengthened the life of the individual with CF. They show that needs vary from individual to individual. Not all treatments work for every person with CF. Various treatments work better for adults versus children. A study of treatments was done to determine what works well and what does not.

*Quality of Life measurement.* It is important to understand the quality of life of each individual with CF because each person with CF has different experiences. Quittner, Buu, Messer, Modi and Watrous (2005) developed The Cystic Fibrosis Questionnaire (CFQ) that is specific to the disease CF. It measures the quality of life related to health in persons with CF who are 14 to 53 years of age. The questionnaire concluded that the young individuals with CF had a better quality of life than those who were older. Those who were older scored lower than the younger adults that participated. There were also other correlations such as “better nutritional status was positively correlated with several weight-related scales. And the measure differentiated between individuals with varying levels of disease severity” (Quittner et al., 2005, p. 2347). The higher score indicated a better quality of life. Two hundred and twelve people with CF participated in this questionnaire over a 10-14 day interval. Similar percents of women compared to men were included in this study. This scale proved to have good reliability on a Cronbach scale with a range of $\alpha = 0.67$ to 0.94 and “test-retest stability was acceptable” on a scale of $r_s=0.45$ to 0.90 (Quittner et al., 2005, p. 2347). The scale’s validity was also tested by “examining relationships between the CFQ, age, pulmonary function, and body mass index” (Quittner et al., 2005, p. 2347). It also serves as a good resource for nurses and other healthcare providers because it shows that as a person with CF ages, his or her needs change. For example, they are more susceptible to weight-loss and decreased body
function than young adults. Nurses should be aware of the changes that occur as CF progresses so that new goals and interventions can be made.

Survivorship model. A model created by Liou, Adler, FitzSimmons, Cahill, Hibbs, and Marshall (2001) concluded that eight characteristics of CF were identified that affect the survival rate of the individual. The characteristics include the following: weight-for-age, gender, whether the individual has pancreatic insufficiency, diabetes mellitus, *Burkerholderia cepacia* infection, *Staphylococcus aureus* infection, and amount of pulmonary exacerbations each year. It shows that not one specific clinical manifestation of CF can be focused on to slow progression. The model was developed to “help researchers and clinicians to evaluate therapies, improve the design of prospective studies, monitor practice patterns, counsel individual patients, and determine the best candidates for lung transplantation” (Liou et al., 2001, p. 345.). One purpose of this model is also to give a guideline for assessing the effectiveness of future therapies (Liou et al., 2001). This model is a 5-year survivorship model, and it analyzes the clinical features of CF. Persons with CF were selected from the Cystic Fibrosis Foundation Patient Registry (CFFPR) which contains longitudinal data on 27,849 people with CF. The model did not include all of the people in the registry; the designers of the model “developed multivariate logistic regression models by using data on 5,820 patients randomly selected from 11,630 in the CFFPR in 1993” (Liou et al., 2001, p. 345). The survivorship model included “age, forced expiratory volume in 1 second as a percentage of predicted normal, gender, weight-for-age z score, pancreatic sufficiency, diabetes mellitus, *Staphylococcus aureus* infection, *Burkerholderia cepacia* infection, and annual number of acute pulmonary exacerbations” (Liou et al., 2001, p. 345).
Lived Experience

Treatments given by healthcare providers may be too complicated or time-consuming for the individual to keep up with; therefore it is important to understand the lives of the individual before implementing treatment plans. The lived experience of different age groups as well as parents of a child with CF will be addressed.

Teenage lived experience. This study concluded that a level of non-compliance is normal for young adults with CF. Their priorities differ from those of healthcare providers. Healthcare providers want each person with CF to follow every possible treatment because their main goal is to slow progression of the disease. Young people with CF have a goal to slow progression as well, but they also have a goal to live their lives as normally as possible. Nurses as well as others need to understand why it is hard for people to adhere to treatments, and it is important to assess how they can help to make the process easier for the person to adhere to (Badlan, 2006). Badlan (2006) explored the experiences of young people with CF and how their experiences impact their ability to keep up with treatments. Thirty-one young adults were interviewed in this study and were analyzed using interpretative phenomenology. Understanding the views of treatment of young people with CF is very important. In addition to this understanding, it is also important to understand the views of parents with young children with CF.

Lived experience of parents. Parents who have children with CF can have many stressful responsibilities. Foster, et al., (2001) interviewed mothers, patients, and siblings in order to assess the lived experience for all of those involved in CF family care. The results of the study returned with all of the parents reporting that they had to remind their children to keep up on the treatments, including parents of older children. Several reasons
were given by parents that described why their children were not adhering to their
treatment plans. These reasons included loss of motivation due to shortened life
expectancy, lengthy treatment time, and resenting the need for having to follow
treatments when their friends have no treatments to follow. This study concluded that
almost all parents are involved in their children’s treatment, even if their children were in
teenage years and that parents were more likely to be involved than the siblings. The
interviews conducted were semi-structured and focused on “management of illness (e.g.,
different forms of treatment, difficulties encountered); the impact of the illness on the
family (e.g., what relationships were like, what happened when the patient was ill)”
(Foster et al., 2001, p. 3). This study also assessed the amount of parental involvement in
their child’s therapy. This study has several limitations. The study involved a small
number of people with differences of ages between sibling and child with CF. However,
this study did conclude that new ways of encouraging adherence need to be developed
because without adherence to treatments, the disease process cannot be slowed (Foster et
al., 2001).

Most children with CF are diagnosed by age two, which puts complete
responsibility on the parents to implement treatments. This can be difficult for parents
and cause stress between couples as well as stress between parents and children. Siblings
of an individual with CF can feel that the CF treatments are the number one priority of
the parents because the treatments are crucial to slowing the progressions of CF. This
may cause the siblings to receive less attention and feel as if they are not an equal priority
to the parents as the child with CF (Berge & Patterson, 2004). In addition to siblings’
feeling as if they are second priority, couples can have strains between each other.
Mothers stated that they took the primary role in treatment for the child with CF and stated that the father was not around to assist with treatments during the day (Quittner, Espelage, Opipari, Carter, Eid, & Eigen, 1998). Fathers stated they have difficulty with attachment to the child with CF as well are more stress related to finances compared to mothers (Quittner et al., 1998).

Young children lived experience. In addition to understanding the lives of individuals with CF, it is also important to understand the coping strategies used by those individuals. A study by Pendleton, Cavalli, Pargament, and Nasr (2002) was composed to assess the importance and significance of religion or spirituality in coping. This study concluded that it is necessary for healthcare providers to assess if there are spiritual/coping needs of the individual with CF as well as their families because many individuals with CF use spirituality as a coping mechanism. This study included 23 children ages 5 to 12 years of age and their parents and conducted interviews with them. One child, a seven-year-old female, wrote, “Jesus is ‘taller than me’ and ‘helps me get better when I am sick’” (Pendleton et al., 2002, p. 3). Other children wrote similar statements about God and being sick; some also drew pictures of themselves with family and God. As a result of this study, it is apparent that more research is needed to determine a more complete understanding of how religion or spirituality is used in coping with CF. Several steps were taken to ensure “trustworthiness” of the research which they compared to standards held for validity and reliability. The individuals chosen for interviews were chosen through a convenience sample in an ambulatory CF clinic. The design of the study was a focused ethnography that “uses techniques such as participant observation and in-depth interviewing to generate, rather than test, hypotheses” (Pendleton et al.,
The interviews were held at the University of Michigan Cystic Fibrosis Center. To increase the trustworthiness, several measures were taken which “included 1) data triangulation (using both child and parent as sources of data), 2) investigator triangulation (independent development of coding systems by 2 investigators), and 3) methodological triangulation (use of several methods to study the phenomenon)” (Pendleton et al., 2002, p. 3). This study has limitations because it only contained a small number of interviews. Although this study included only a small sample, it is helpful for nurses to increase their understanding of spirituality in coping and also in treatment plans. For some individuals, praying is an intervention that they use to help cope with the disease, and some individuals also believe that praying will help find a cure for their disease.
Chapter III
Methodology

*Phenomenology Method*

The purpose of this research was to determine how CF affected the lives of those who have inherited it as well as the family members and loved ones involved. By doing this; healthcare providers as well as others will be able to more fully understand the effects of CF on everyday life.

Phenomenology method was used because “phenomenology is both a philosophy and a research method that explore and describe everyday experiences in order to generate and enhance the understanding of what it means to be human” (Russell, 2004, p. 221). The purpose of this method of research was to describe the lived experience of someone with a chronic illness. The lived experience is part of a “holistic model that reflects the interrelationship of body, mind, spirit, and environmental” (Russell, 2004, p. 221). This explored what it means to be human and what experiences human beings had living with CF.

Living with a chronic illness is just that; it is a lived experience. This method was chosen to help attain information to enhance the understanding of the lived experience of people with CF. CF is a disease that must be treated for a lifetime and requires treatment plans to become a way of life.

*Avoiding Bias*

Phenomenology included bracketing to help avoid any biases. Bracketing is described as “identification of any previous knowledge, ideas, or beliefs about the phenomenon under investigation” (Russell, 2004, p. 219). It was important to view
anyone who has a chronic disease, such as CF, as a whole and not just categorize him or her through the disease.

Sample

All participants were English speaking. One male and two females were interviewed. In addition to persons with CF, a parent who has a child with CF was also interviewed. A total of three individuals were interviewed. Individuals interviewed were from various states including, Washington, Oregon, Idaho, Montana, and Illinios.

Confidentiality

All information was kept in a locked cabinet. No names or identifiable information was used in research. Informed consent was gathered for each individual who volunteered to be interviewed. For children interviewed under the age of 18, an informed consent was signed by a parent or guardian. Informed consent was given by parents for all young children used. Pseudo names may be used.

Data Collected and Setting

Information was collected through personal interviews. A comfortable location was chosen for each interview. Audiotape was used for each interview and was transcribed. Interview questions included questions about how CF affects everyday life and what is it like to live with CF.

Study Limitations

This study contained a small sample. These findings cannot be generalized to all individuals with cystic fibrosis.
Data Analysis

Colaizzi’s Method was used to conduct and analyze these interviews. These are described as “lengthy and repeated interviews to facilitate full description” (Fain, 2004, p. 231). The data were collected through participant subjective accounts of the lived experience and significant statements were extracted and put into clusters. This method served as a structure for interviews and data collection.

Colaizzi’s Method

1. Descriptions of the lived experiences
2. Diverse collected descriptions of the lived experience
3. Aggregated significant meanings and findings into clusters and important themes
4. A written exhaustive description
5. Return interviews with the participants for validation
6. Incorporated new data into a final very exhaustive description

(Fain, 1999, p. 230)

Rigor

To ensure integrity of the study steps of analysis method were followed. Methods and result were continually coded with faculty and results were supported by personal quotes obtained through interviews.
Chapter IV

Results

What we learn from textbooks and seminars is great, but patients are a nurse’s greatest teachers. I’ve been honored to share in the lives and sometimes the deaths of young people after lung transplants and they’ve ignited my passion for nursing in a way I’d never dreamed possible (Whitlow, 2005, p. 32).

The purpose of this thesis was to gain insight into the lives of individuals living with CF. Five interviews were conducted which resulted in several themes. These themes included utilizing family support as a coping strategy, time-consuming treatments, decreasing energy, and staying positive. Each theme has supporting information from interviews and quotes that give validation. Each individual was given a pseudo-name in order to maintain privacy for all personal information shared.

Utilizing Family Support as a Coping Strategy

All participants expressed the need to develop coping strategies in order to have the strength to continue treatments and maintain a positive attitude. The main coping strategy used by all participants was family support. Having a support system that cared for them and understood their disease and all of the difficulties that accompanied it was essential.

Richard discussed how it was difficult for him to get back into his routines after being in the hospital. He stated “Family support really helped me recover from the hospital.” His fiancée and parents were his main source of support that he utilized. He discussed that just having someone there who understood his disease and his needs helped him to cope with CF and gave him strength to keep himself healthy.

Mary talked about how CF was not one dimensional; she stated, “It’s hard to nail it down to one thing: its psychological, it’s emotional and its physical, it’s all those things
rolled into one, and at the same time you are just trying to be a normal person.” She expressed that because CF affects all aspects of a person, it is essential to find a support group whether it is family or friends or anything else. She talked about how her parents were her support system and without them she would not be here today. Her parents are also very involved in CF fundraising, creating their own fundraiser in their daughter’s name.

Nancy expressed that she utilizes her mother a lot as a support person. She recalled one time when she was a young teen that her mother rushed her to the emergency room at a children’s hospital in the middle of the night. She ended up having a her first really bad lung infection at that time and expressed that she was so happy to have her mother there with her. She said, “My mom helps me out a lot; my mom has been the best.”

Claire, the mother of a four year-old daughter with CF, expressed that she needs family when her stress level gets high. She talked about the need for having breaks from a hectic lifestyle of raising a child with CF and also raising several other young children as well. Having a break, such as getting a babysitter, or leaving the children with a family member, allowed her to have time to herself and therefore be able to deliver better care to her children.

*Time-consuming Treatments*

The most time-consuming treatment expressed by all of the participants was the need to do breathing treatments and airway clearance. The participants’ treatments ranged from doing them twice a day to three or four times a day, depending on the severity of their CF and if he or she had a current infection at the time. Each breathing treatment
takes an average of a half an hour, most consisting of the use of a nebulizer to administer a bronchodilator, such as Albuterol, followed by the use of a decongestant, such as Dornase. Three out of the five participants said that they used a vibrating vest for airway clearance while they did breathing treatments so that they would not have to take the time to do airway clearance in addition to breathing treatments. Two out of five participants did not have a vibrating vest and had to do airway clearance separately from their breathing treatments. All participants stated that the need for a routine was necessary in planning for fitting the treatments into their daily schedules. All also expressed that the treatments were time-consuming and could become quite burdensome.

When asked how they were able to get through the treatments, Richard stated, "It’s a routine and sometimes I have to remind myself that I need to keep doing it and that there is a reason that I am doing it, but it definitely can get old doing it every single day.” Mary expressed that CF treatments become a part of life and that it is just something that is necessary for her to stay alive. She stated, “A lot of people say that CF does not run their life, but it does if you take care of yourself and are really putting in the time and the effort.”

Kyle explained that he does not use any conventional ways of airway clearance, such as the use of a vest or chest physiotherapy. When asked how he gets good clearance he said, “If I want to do airway clearance I get on my snowmobile and ride, and within about ten minutes I need to stop because I have so much clearance of mucous and everything else.” He said that if he stays active and works outside, he is able to clear out his lungs and does not need to use any of the conventional methods that are recommended to him.
Claire was in the process of getting a vest for her child with CF, but was having difficulty convincing her insurance company to pay for it. The insurance company claimed that it should not have to pay for it because there is an alternative way of airway clearance using manual percussion and postural drainage. Claire expressed frustration with this because what the insurance company did not understand was that the vest is much faster and can be done while her daughter does breathing treatments.

When Nancy was asked, if her treatments affect her daily routine, she answered. “No, my treatments do not ever affect my daily routine, because that pretty much is my routine daily; if I didn’t do them I wouldn’t be here.” She went on to explain that she has always had to do them and they have always been a routine in her life so she does not know any other routine that does not include her daily treatments. She expressed that they do take up a lot of time and they can be discouraging, but she does them because she has to do them.

In addition to time-consuming breathing treatments, several individuals expressed the difficulty of gaining weight and maintaining that weight. Individuals with CF are required to take supplemental digestive enzymes that their pancreas does not produce. They must take these enzymes before every meal and every snack. Richard stated, “Everything gets tiring at times; sometimes I want to just eat a meal without having to take enzymes, or if I get home late at night and still have to do my breathing treatments, sometimes I want to go to bed, and I am not going to lie. Sometimes I just skip them and go to bed.”
Decreasing Energy

Decreasing energy was one of the main discouraging aspects of CF according to the interviews conducted, especially in adults with CF. It was discussed by several participants that once a person has to quit a job due to symptoms of CF, he or she has to search for other ways to define himself or herself as a person. He or she can no longer answer the question, what do you do? The answer may just simply be: “trying to live.” Participants expressed that some people perceive them as lazy when they do not have a job or have to take time off work.

Mary stated, “I see my other adult friends with CF struggle, and the day that they have to stop working is one of the hardest days because we define ourselves by what we do, not by who we are.” Having decreased energy with CF becomes a chronic problem, so many adults with CF have to quit their jobs in order to have energy to do their treatments and stay healthy. As evidenced by the quote above, having to quit a job due to CF is a very significant, emotional problem.

Richard expressed frustration with his decreasing energy when stating,

When I was younger and when I was in high school I could pretty much do whatever I wanted and keep up with my friends and people my age, but now there is definitely less energy, my lungs are not what they used to be.

He explained that when he was a child, he was able to play all of the sports he wanted to play and did not feel different from the other children. When he graduated from high school, he was admitted to the hospital for the first time; this was when he said he started to notice a decline in his lung function.

Nancy also said that as a child she had much more energy than she has now. As a child she could do whatever she wanted but now she says that “everything is weakening;
my lungs are weakening, sometimes now I’m to the point where I can’t even get out of bed to take a shower.” Nancy was in college and had to drop out due the negative effects of CF.

Claire expressed that she already noticed some differences in energy between her child with CF and her other children who do not have CF. Claire explained that her child still wants to participate in all of the same activities as the other children, but,

If she is playing hard then she will start coughing more, and we will have her sit down for a while and have a glass of water and that seems to calm her down and make her coughing go down. I think it is because her mucous gets too thick, and the glass of water seem to help with that as well.

Decreasing energy in CF can not only cause adults to have to quit work, teenagers to quit school, or young children to sit out in activities, but it also affects whether the individual has enough strength to continue time-consuming treatments.

Staying Positive

Through personal interviews, it was made clear that all of those who were interviewed expressed that CF has many burdens, but surprisingly all of them also said that they find ways to stay positive. Each individual described different ways that he or she looks at life in order to stay positive regardless of having a debilitating illness.

Mary described having CF as a blessing. She said, “In a way CF is a blessing because even though I will only be here a short time I am going to learn a lot more in that short time than some people learn in a lifetime.” When Mary was asked how she was able to stay positive she simply stated, “I guess you could say that my religion is positivity. It’s true I try to find positive in every negative.” She went on to explain that having CF is a negative, but by having CF she is able to appreciate life more fully and live in the
Like other individuals interviewed she expressed the importance of spreading awareness and raising money to help support CF research.

Richard expressed, “I know sometimes I wish that I didn’t have CF, but at the same time CF has shaped me into who I am.” He described that he did not know any other way of life besides living a life with CF.

Nancy said that sometimes it is really hard for her to stay positive. She said that the way she stayed positive was to “suck in every moment.” She expressed some anger by stating, “I believe in God, I do, but I am mad at him. I’m wondering why I’m here suffering, what’s my whole point.” She went on to express that one way she stays positive is to open her friends’ eyes to what life is all about. She believes that she has helped to teach them not to take anything for granted and to live life to the fullest.

Several individuals interviewed expressed that one way they were able to stay positive was to have hope. They all expressed that they go through hard times due to CF and that they are not able to stay positive all the time, but they all utilize coping strategies, complete time-consuming treatments, and do everything they can despite their decreased energy. Ultimately they all have hope because they all believed that research for CF is getting closer to finding a cure.
Chapter V
Discussion

Utilizing Family Support as a Coping Strategy

Utilizing support systems, specifically family, was the main coping strategy used by all participants interviewed in this thesis. Siems (2007), she explained her role as a parent of a child with CF. Siems expressed her feelings of fear and sorrow and said that she was able to cope with her daughter having CF by utilizing her resources of family support, forming a team with health care workers, and becoming involved with the Cystic Fibrosis Foundation (CFF). She shared, “It constantly awes me to see the contributions others are willing to make, even when they have no direct connection to CF. It assures me that we are not alone in our struggles” (Siems, 2007, p. 429). As a mother of a young child with CF, she explained that she felt compelled to not only physically care for her daughter to the best of her ability, but to also become involved in the CFF and contribute to CF research. In referencing all of her responsibilities as a mother she stated, “It would be impossible for me to do this work without the support of my family and friends” (Siems, 2007, p. 429).

One article focused on the increased need for individuals with CF to utilize family support when transitioning from teenage years to adulthood. Children with CF depend on family a great deal, and as they mature and eventually reach the age of adulthood, they should become more independent with their treatments and more likely to succeed living on their own. This article explains the family role in helping to prepare young people with CF to transition into adulthood: “Family connectedness, family role models, family concern for the well-being of the child, and autonomy at home are all factors identified
that foster resilience in young people” (Hink & Schellhase, 2006, p. 261). Transitioning into adulthood can be difficult for all individuals. Hink and Schellhase recommended that an individual with CF may need more family support during this period than someone not living with CF. The individual with CF may have fears regarding personal responsibility and independence from family members, such as parents. Therefore, there is also great importance in “encouraging a good relationship with the adult CF team and involving both pediatric and adult teams along with the family in the development and implementation of a transition program are both important factors in successful transition programs” (Hink & Schellhase, 2006, p. 262).

*Time-consuming Treatments*

As expressed by all individuals interviewed, time-consuming treatments include breathing treatments, chest physiotherapy, and pancreatic enzyme replacements. All participants expressed that breathing treatments were the most time-consuming and all expressed that treatments can be burdensome. A qualitative study researching the lived experience of growing up with CF stated, “A curative treatment is not yet available. Hence, the patients have to go through complex and time-consuming therapeutic regimes consisting of chest physiotherapy, inhalation therapy, pancreatic enzyme supplements, and regular antibiotic therapy” (Gjengedal, Rustoen, Wahl & Hanestad, 2003, p. 150). The study also researched if children with CF felt normal or if they felt different from other children without CF. Most children did not seem to feel any different than other children. One mother expressed why she thought this was true by stating, “Even if the everyday life of their children is different from that of well children, the CF children do not experience it as different: to them this life is quite normal” (Gjengedal et al., 2003, p.
Several mothers involved in this study said that they felt their young children didn’t feel any different from other children because they simply did not know any other way of life. This changed greatly as children approached adolescence. During the adolescent period is a time where more respiratory problems and lung infections can manifest. As a result the individual is required to do more therapy, including inhalation therapy and antibiotic therapy. Treatments during this period are more time-consuming resulting in the adolescent feeling different from his or her peers.

*Decreasing Energy*

Decreasing energy was one of the main complaints of all individuals interviewed. This became even more relevant if the individual was in adulthood. In the study written by Gjengedal, Rustoen, Wahl, and Hanestad (2003), time-consuming treatments were related to decreasing energy in individuals with CF. The study explained that most individuals in the study they conducted wanted to follow all of the therapeutic regimens and continued to keep with the treatments because they knew that they needed them to stay healthy and expressed that they did the treatment “simply because the therapy makes them feel better” (Gjengedal et al., 2003, p. 154). Although most expressed that they knew they needed the treatments, they also expressed that the treatments consumed a lot of their energy. The study concluded that “most of the informants complained about loss of energy and urgent need for sleep. In addition, as the disease progresses, the frequent lung infections produced symptoms such as uncomfortable coughing and breathlessness, and increases the feeling of being exhausted” (Gjengedal et al., 2003, p. 154). This study expressed that there was a strong relationship between time-consuming treatments and
decreasing energy leading to some feelings of exhaustion. As the individual with CF ages, there is an increase in treatments needed and therefore a decrease in energy levels.

Staying Positive

After conducting interviews and researching CF, it is apparent that CF has many burdens and can cause fear and anger in those who are affected. Despite all of the negative aspects that come with having CF, all individuals expressed the need to stay positive and some expressed that staying positive included finding a source of hope. In a study conducted by Abbott, Dodd, Gee, and Webb (2001), coping strategies were compared with treatment adherence. The study concluded that “higher levels of optimism and self efficacy were correlated with greater treatment adherence in CF adolescence and adults” (Abbott et al., 2001, p. 316). This study reported that this type of coping was considered “active” coping and was generally considered a positive way to cope. It was also reported that an avoidance method of coping including denial was considered the most negative way of coping with CF and was related to non-adherence to treatments. Included in the “active” coping method was talking with family or friends and having hope.

Nursing Recommendations

Future research is needed to explore and understand the lived experience of those with CF. The psychological impact of CF is not well understood. Psychological issues include: fatigue, infertility in males, and depression.

Conclusion

Living with CF has been proven to be difficult and burdensome. Individuals with CF are required to do time-consuming and energy-consuming treatments in order to stay
healthy and prolong their life expectancy. The purpose of this thesis was to gain insight into the lives of individuals living with CF. By conducting personal interviews and researching the lives of those with CF, nurses and other healthcare providers can begin to learn how to care for an individual with CF. Having a family support system and finding ways to stay positive are two ways that individuals with CF find the strength to continue treatments and live a life with CF. By researching the themes in this thesis nurses and other healthcare providers need to have an understanding of what it is like to live with CF in order to care for each CF individual in a holistic manner.
References


Cystic Fibrosis Services Pharmacy Educational Series. (2007). The role pancreatic enzymes play in achieving good nutrition.


