The Family Experience of Living with Cystic Fibrosis:
A Case Study of Two Families

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Abstract

The diagnosis of Cystic Fibrosis (CF) affects not only the well-being of the individual with the condition, but also the well-being of other family members and the family as a whole. Our understanding of the family experience of living with CF is limited. Much of the existing research has focused on the experiences of individual family members, rather than the family as a whole. Therefore, the purpose of this study was to explore the family experience of living with a child that has CF. The guiding framework for the study was the Resiliency Model of Family Stress, Adjustment and Adaptation. Semi-structured interviews were conducted with parents from two families living with CF. Findings from this study illustrate how important it is for nurses and other health care providers to recognize that no two families living with CF have the exact same experience. Each family goes through their own unique journey. How a family responds to the ongoing challenges associated with raising a child with CF will depend on factors such as age of the child, number of children, work status of parents and family variables like family demands, family appraisal, family resources, and family problem-solving and coping. Nurses who understand this will provide a better quality of care, which will lead to greater patient and family satisfaction, as well as increased job satisfaction for the nurse.
Cystic Fibrosis affects approximately 30,000 individuals in the United States and 70,000 worldwide (Cystic Fibrosis Foundation, 2014). The diagnosis of CF affects not only the well-being of the individual with the condition, but also the well-being of other family members and the family as a whole. Our understanding of the family experience of living with cystic fibrosis continues to be rather limited. Much of the existing research has focused on the experiences of individual family members (Besier & Goldbeck, 2011; Driscoll et al., 2010; O’Haver et al., 2010; Wong & Heriot, 2008), rather than the family as a whole. Another line of research has been the parent-child transition in managing cystic fibrosis (Leeman, Sandelowski, Havill, & Knafl, 2015). Minimal attention has been paid to how family factors influence adaptation at the individual and family level. Therefore, the purpose of this study was to explore the family experience of living with a child that has cystic fibrosis.

**Background**

Cystic Fibrosis (CF) is found to be one of the most common autosomal recessive, life-limiting diseases in Caucasians (Ernst, Johnson, & Stark, 2010). This autosomal recessive disorder results from mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene. The CFTR protein generates a chloride channel present in the apical membrane of epithelial cells throughout the body (Griesenbach & Alton, 2015). The mutations in CFTR gene alter chloride transport across membranes throughout the body. This causes a compensatory influx of sodium to maintain electro-neutrality and a consequent influx of water, extracellular de-hydration, and formation of thick mucous in the airways (Marson, Hortencio, Aguiar, & Ribeiro, 2015). The thickness of the mucous in the airways causes problems with mucous clearance, which subsequently leads to lung inflammation and infections.
There have been nearly 2,000 mutations of the CFTR gene identified in 2014 and information about these specific mutations has provided knowledge for the genotypic and phenotypic characteristics of CF (Marson, et al., 2015). Clinical characteristics are different with each mutation and with each individual. Nearly every individual with CF experiences respiratory symptoms with gastrointestinal symptoms being the second most common. Gastrointestinal consequences include exocrine pancreatic insufficiency (in an estimated 90% of individuals), which sets the stage for CF-related diabetes in an estimated 50% of adults over the age of 30 years old (Ernst, Johnson, & Lark, 2010, p.263). Other gastrointestinal effects can include poor nutrient absorption (especially for fat), biliary cirrhosis, bile duct proliferation and excessive absorption of fluid, increasing the risk for intestine obstruction (Ernst, Johnson, & Lark, 2010, p.264). Because respiratory and gastrointestinal symptoms are the most common, early symptoms that are indicative of CF are respiratory infections and failure to thrive. Unfortunately, respiratory symptoms and gastrointestinal symptoms are not the only clinical characteristics of CF. Additional symptoms that occur across the lifespan include chronic sinusitis, nasal polyps, and late onset puberty (Ernst, Johnson, & Lark, 2010). Infertility is present for nearly 95% of males due to the absence of the vas deferens and for 20% of females due to ion transport issues in the genitourinary system, such as abnormal cervical mucous (Boucher, 2008, p.1632).

Over the past few decades there has been a substantial increase in the treatments available for individuals with CF. There have been significant advances in both inhalation medications and the delivery devices. Intelligent nebulizers and dry powders have become the commonplace for CF treatment (Agent & Parrott, 2015). Inhaled medicines generate high levels of a drug within the airways with limited systemic effects, offering safe and convenient antibiotic and mucolytic therapy for individuals with CF (Agent & Parrott, 2015, p.110). There also been an increase in
use of high frequency chest wall oscillation (HFCWO) over chest physiotherapy. HFCWO assist devices generate positive or negative respiratory pressures excursions to produce high-frequency, small-volume oscillations in the airways, which generate coughing and mucous mobilization (Nicolini, et al., 2013). This increase in use of HFCWO could be attributed to the portability of using this mechanical form of chest physiotherapy. HFCWO usually includes an inflatable vest connected by tubes to a small air-pulse generator and manufacturers, such as Respirtech and Electromed. These have made the systems portable and easy to use for patient convenience. This increase in treatments and the increase in availability of genetic testing, newborn screening, and the opening of more CF Foundation-accredited care centers have all contributed to the longevity and enhanced quality of life for individuals with CF. The median predicted age of survival for individuals with CF has increased from 33.4 years in 2003 to 40.7 years and 2,697 individuals with CF were 40 years old or older in 2013 (Cystic Fibrosis Patient Registry Data, 2013). In 1986, 29.2% of individuals with CF were 18 years old and older and in 2013, that has in increased to 49.7% (Cystic Fibrosis Patient Registry Data, 2013).

As the life expectancy for individuals with CF increases, so do the type of demands placed on families. Individuals with CF are now experiencing a wide range of life experiences such as going to college, getting married and having children. While this is exciting, it also places different demands on their families and the demands will continue for a longer period of time (Foster et al., 2001). For example, when individuals with CF have children, there is a good chance they may not live until their children reach adulthood. Because of this, they may need to rely on their family to assume additional caretaking demands. (Foster et al., 2001).

Because individuals with CF are in and out of the hospital year after year, healthcare providers become an integral part of their lives for decades. Years ago, most individuals with CF
were cared for in pediatric hospitals and specialty clinics for children with CF. Now that individuals with CF are living longer, their care is being transitioned to health care providers who care for adults. Therefore, it is essential that all health care providers, not just health care providers in pediatrics, have a good understanding of not only care needs of individuals with CF, but the role of the family. Health care providers who have a good understanding of how families influence, and are influenced by, the experience of living with CF will be better prepared to work with the family. This in turn, will improve not only the quality of care, but patient and family satisfaction with care. It may also improve job satisfaction for the health care providers working with them.

While there has been limited research on how family factors influence adaptation at the individual level, there has been some research on the quality of healthcare delivery for not only individuals with CF but their families as well. The CF Foundation developed an experience of care survey to systematically collect and learn directly from individuals with CF and families about their impressions and observations of CF health care delivery (Homa, Sabadosa, Marrow, & Marshall, 2015). An improvement opportunity that was identified was the need to assess the mental health of the individual with CF and the family. This testifies to how important it is to perform more research on the family unit and not just the individual.

**Purpose and Objective**

As noted previously, the purpose of this study was to explore the family experience of living with a child that has cystic fibrosis. The case study method was used to compare and contrast the life experience of two different families and how each family unit adapts to the daily stressors and challenges associated with having a family member with cystic fibrosis. The
Resiliency Model of Family Stress, Adjustment, and Adaptation was the guiding framework for this study.

**Theory**

The Resiliency Model of Family Stress, Adjustment and Adaptation looks at the whole family and their ability to face the challenges of a new stressor, such as an illness or disorder, and to adjust and adapt to the pileup of demands that take place over time (McCubbin & McCubbin, 1993). The adjustment phase of the Resiliency Model relates to minor challenges the family may face and the ability of the family to continue with its current way of functioning without much interruption (Friedman, Bowden, & Jones, 2003). The adaptation phase of the Resiliency Model involves the family facing a major stress where the family must take on a series of changes in order to adapt to the stress (McCubbin & McCubbin, 1993). The components of the adaptation phase are pileup of family demands, family and situational appraisal, problem solving and coping skills, and family resources (McCubbin and McCubbin, 1993; Robinson, 1997).

*Pile up of Family Demands*

Families often times have stressors and can have multiple stressors at the same time. Stressors in families with children with a chronic illness can be more long-term and can have an additive effect. This additive effect of pre-existing stressors, current stressors, and future stressors of these families contributes to the pile up of family demands. Six broad categories of stressors and strains are identified in the Resiliency Model: (1) The stressor and related difficulties; (2) typical transitions in individual family members and the family as a unit; (3) preexisting family strains that accumulated over time; (4) situational obligations; (5)
repercussions of the family efforts to cope; (6) ambiguity within and outside the family (McCubbin & McCubbin, 1993; Friedman, Bowden, & Jones, 2003).

Family Appraisal

Family Appraisal refers to the family’s subjective definition of the illness and the difficulties related to the illness (McCubbin & McCubbin, 1993). The family appraisal can reveal several things, including the family’s view of how difficult the disease and related hardships are, the capacity of the family to manage the situation, and the appraisal can predict how well the family will adapt (Friedman, Bowden, & Jones, 2003; Robinson, 1997). Families who have a more positive outlook and attitude towards the illness and its difficulties are more likely to cope and adapt easier than families who have a negative outlook. Situational appraisal is a facet of the family appraisal and shows how the family manages the demands related to the illness. The family’s situational appraisal discloses the sufficiency or insufficiency of the family to manage the situation (Friedman, Bowden, & Jones, 2003).

Family Problem Solving and Coping Skills

Problem Solving refers to the family’s ability to sort a stressor into manageable portions, to recognize alternative ways to deal with each portion, to begin steps to resolve the particular issues, and to form and refine communication necessary to the production of problem-solving efforts (McCubbin & McCubbin, 1993). Coping skills refer to how family members manage a situation, including how they preserve or strengthen the family as a unit, uphold the emotional health and wellness of its members, acquire or utilize resources, and begin efforts to resolve family difficulties produced by a stressor (McCubbin & McCubbin, 1993). Problem solving and coping skills are important for any family to work through a stressor or a crisis but they are
especially important skills to investigate in families who live with a chronic illness to assess if the family is well adapted or if there is maladaptation.

*Family Resources*

Family resources pertain to the competence and capacity of the family to deal with a stressor and its conditions by hindering the situation from becoming a crisis or creating disturbance in established patterns of family functioning (McCubbin & McCubbin, 1993). McCubbin & McCubbin identify three different types of resources: (1) the individual family members; (2) the family working as a whole; (3) the community. Family resources can encompass a wide variety of assets. Resources can include, but are not limited to, finances, family support, access to medical care, intelligence, community support, and specific personality traits.

*Family Adaptation*

Family adaptation is the desired outcome of the Resiliency Model. The adaptation phase is a process that begins with a critical event or stressor. This is a life event that impacts a family member and produces or has the potential to produce changes within the family system. How the family goes through this process of adaptation depends upon the pileup of the demands, family appraisal, problem solving and coping skills, and family resources. Adaptation is a process in which families react to the demands of a stressor and make changes that are necessary for the coherence and well-being of the individual family members and the family as a unit (McCubbin & McCubbin, 1993). While a well-adapted outcome is optimal, there can also be a process of maladjustment. Maladjustment involves disruptions in the family’s firmly incorporated patterns and usually results in a state of crisis and is characterized by disorder, disorganization, and an inability to reestablish family stability (McCubbin & McCubbin, 1993).
Families of children with CF can encounter multiple stressors such as difficulty with feeding, managing medications, failure to thrive in the affected child, numerous hospitalizations, and keeping up with the rigorous treatment schedule. These stressors can significantly affect the family unit and their ability to function. It is important for the family to adapt to these stressors and to be able to make changes in their normal family functioning. The Resiliency Model is a guiding framework that can help show how families move through the adaptation process.

**Methods**

*Design*

This study was a descriptive, comparative case study of two families of children with CF. By conducting in-depth interviews with families, the investigators were able to obtain a detailed understanding of what life is like living with a child who has CF. A qualitative method was used so that the families were able to give a detailed explanation of their story from their own viewpoint.

*Setting*

The families were interviewed at a location of their choice. Family A was interviewed in the apartment clubhouse of the main investigator. Family B was interviewed in their own home. Even though the settings were not identical for each family interview, both settings allowed for a quiet space that was conducive for conducting interviews.

*Sample*

The study sample included two families. Both of these families were recruited to be part of a larger study conducted by Dr. Marcia Van Riper, *A Family Experience of Living with Cystic Fibrosis*. In order to participate in the study, participants had to have at least one child with CF.
Family A consisted of one child with CF, two children without CF, and a mother and father. Family B consisted of one child with CF, one child without CF, and a mother and father.

**Data Collection Procedures**

Families were informed of the study by either the main investigator or Dr. Van Riper. They received a description of the study and an overview of the type of questions that could be asked during the interview. Once they expressed interest in participating an interview was set up at a time and place that was convenient to them.

The study involved voluntary participation and both families were told that they did not have to answer any questions that they did not feel comfortable answering. Both families were also told that they could withdraw from the study at any point without negative consequences. Full disclosure of the study was provided to the families and the study did not involve any risk. Fictitious names are used in the results section of this study.

**Measures**

An interview guide previously used by Dr. Van Riper (Appendix A) in interviews she has conducted with families with CF was used to structure the interview questions. The interviews consisted of open-ended questions centered on a) how the family member defines the experience of having a child with CF b) what kinds of stressors related to their child having CF they have experienced, c) how they have adapted to the challenges that have arisen, d) what and/or who has influenced the ways they manage the challenges, and e) what/who has helped them cope during the stressful times.

**Data Analysis**

Both interviews were recorded on a digital recorder. Then, they downloaded on one of the investigator’s computers. The data was then organized into time segments and categorized
into the components of the Resiliency Model of Family Stress, Adjustment, and Adaptation. The Resiliency Model components were identified for each family separately and then compared to show how the families were adapting to having a child with CF. The categories used are pileup of family demands, family appraisal, problem solving and coping skills, and family resources. The data was summarized by discussing how the families adapted to the critical event of having a child with CF.

Results- Family A

Pileup of Demands

*Diagnosis-* Jan (mother) and Richard (father) had a 6-year-old son, Dawson, when their twins, Carter and Walker, were born at 32 weeks. Jan’s water broke and she had an ultrasound to see if she could keep the twins in longer but Walker had an intestinal blockage so an emergency C-section was conducted. Walker immediately had an abdominal surgery to fix the blockage and the surgeon informed Jan and Richard that he had never seen this except for in children with CF. They received the unofficial diagnosis then but the official diagnosis came nine days later. At this time, they did not know anything about CF except that the kids were hospitalized occasionally. Walker and Carter were both hospitalized in the NICU, Carter for three weeks and Walker for about 4 months. Walker also stayed on a children’s floor for a month after the NICU. During this time, Jan had the surgery for her C-section and also had to have her gallbladder removed. This time proved to be exceedingly hard for Jan. She stated,

“I had to depend on lots of other people: babysitters and friends to pick up my other son from school. All of sudden I wasn’t able to do those things for him anymore. That was hard. I started to feel guilty about what I was missing everywhere. Like, if I was home I
felt guilty about leaving Walker. If I was with Walker, I felt guilty about leaving Dawson or Carter.”

In present day, Dawson is 12 and Carter and Walker are 6. Richard and Jen own a restaurant and this is Richard’s full time job. Most of the care falls on Jan because she doesn’t have another job but Richard is still very involved in Walker’s care and is a great support. When asked what the main challenges have been in the last few years, Jen stated, “I think it is the care. The care is so demanding. You don’t get a break, there is no break, right? If you take a break, the disease gets more of a hold.” Although the day to day treatment for Walker is less demanding now than it was the first three years of his life, it is still a lot. He has his daily routine of vest treatments, breathing treatments, and enzymes. The vest alone is done 2-3 times a day. This routine could be overwhelming with just one child but Jan and Richard also have Carter, who plays hockey, lacrosse, and baseball, and Dawson who plays hockey, lacrosse, basketball, and is in band. All three boys are in school full-time and Jan volunteers for several school events. Although Richard does the majority of the work for the restaurant, Jan and Richard both attend events and functions put on by the restaurant several times a year. They are also very active in their local church and attend regular activities there. Jan stated, “It is hard to juggle an illness with healthy people in the family and maintain some sort of regular life as much as you can.”

Family Appraisal

Jan and Richard could not imagine their lives without Walker and know their family is better and stronger for having him be a part of it. Jan has a positive outlook on how fortunate they are to own a business that allows their schedules to be so flexible. She acknowledges that they wouldn’t be able to do as much as they do without that flexibility. She stated,
“We don’t want to be the family that has cystic fibrosis. We want to be a family that happens to have a member who has cystic fibrosis, right? That is the main thing. We don’t want him to be all about cystic fibrosis. It is not his identity. He is Walker and he is a person and he just happens to have this.”

She continued to explain that CF is not the focus of their lives, it is a part of it. Having a viewpoint like this, helps them to keep everything in focus and to fit it in as a part of who they are as a family. After learning of Walker’s diagnosis, both Jan and Richard recommitted to each other that no matter what happens, they are going to be a family. They already have an identity as a family that cannot be broken and CF is just an extra thing. Jan also stated that Walker having CF has deepened her oldest son, Dawson’s, character. He is a lot more compassionate and understanding of people and that there are special times that come out of these hard circumstances.

**Family Resources**

The existing resources that Family A have include: running a successful restaurant business that allows Jan to stay home, good health insurance, access to UNC Cystic Fibrosis specialty clinic, strong and stable marriage, and community support. Jan and Richard also have open communication, immense support from family and friends, and are both very well educated. The entire family is also involved in the Boomer Esiason Foundation and involved in supporting further research for a cure for CF. Jan stated that their faith is something that has carried them through and will continue to do so. They both know that God has a special purpose for Walker and is going to use him to do great things.

Jan’s parents live in the same city and have been a tremendous help and resource to her. They allow her and Richard to get the occasional break from taking care of all three boys at
once. Because of Richard’s job, they have also been able to have several nannies to help with the day-to-day things around the house and with the boys. In the first few years of the twins’ lives, they were able to have a nanny 40 hours a week. Now, they do not need help 40 hours a week but have a few girls who can help out when needed.

Jan’s positive attitude and strength were present throughout the interview. She continues to focus on the positives that have come from CF and does not complain. Even in the midst of the twins first year of life when their world was changing so rapidly and she felt guilty, she managed to have a positive spin on her situation. “I finally started to settle down and decided that I was going to be present where I am because that is what I can give to this child right now. I can’t do anything when I am away from them. So, I started to try to be more mindful of that.” Her love and devotion to her husband and her three boys was very apparent in the interview. She is the backbone of their family and continues to keep everyone on track by organizing all school activities, after school activities, family events, church activities, Walker’s treatments, and occasional hospitalizations.

Family Problem Solving and Coping

Jan and Richard work together when it comes to problem solving in their family. Although Jan does most of the care, they both value each other’s opinion and have equal input. Jan explained, “We consult each other but also take advantage of all services/ideas of the doctors nurses, dieticians, social workers at the hospital. Also professional psychologists to help us navigate the challenges of family life.” A lot of the problem solving comes with the logistics of having to stay in the hospital for multiple nights on occasion. When Walker is hospitalized, most of the time Carter and Dawson do not stay at the hospital. This means that when Jan and Richard switch places at the hospital, one of the kids has to be alone or they have to find
someone to watch Brett at the hospital or watch Carter and Dawson at home. With Jan’s parents
moving close, this process is easier but still difficult.

Jan expressed that her faith is the main way that they cope with things in life. They truly
believe that God has a purpose for everything and everyone and Brett’s story can impact many
other lives. The help they have received from friends and family have also helped them cope
with living with CF, especially in the first year of Walker’s life. Jen explained, “We were
drowning. If it wasn’t for other people helping, I don’t know what we would have done. I don’t
think we had to cook for ourselves for the first few months after the twins were born.”

Jan and Richard both accepted advice from others and actively researched things about
CF. Jan stated that she didn’t research initially because it was too much, but on her own time,
when she was ready, she looked in to everything as she needed it.

Family Adaptation

Walker’s unexpected diagnosis of CF was the critical event and started the process to
adaptation. Family A did move through a lot of emotions during the few weeks after Walker’s
diagnosis but had a strong foundation in their marriage and in their faith that did not waver. They
had existing resources but because of Walker’s medical needs, they needed additional resources.
Family A was slow to start adapting their normal lives to having a child with CF because Walker
spent nearly 5 months in the hospital. Staying and visiting the hospital became their new normal
and when they were finally all under one roof, they quickly adapted and made the necessary
changes to accommodate Walker’s medical needs while making minimal disruptions to their
already established family functioning. Despite the critical event of Walker’s diagnosis, Family
A has accepted Walker’s diagnosis, continued to flourish, and has impacted the lives of others.
Family A has kept a positive outlook and continued to expand and explore the resources
available to them. The cornerstone of this well-adapted family is Jan. She has actively made efforts to manage the stress and demands that comes along with having three boys, one of whom has CF. The adaptations of Family A have strengthened them as a unit and allowed them to function healthily and effectively.

**Results- Family B**

**Pileup of Demands**

*Diagnosis-* Debbie (mother) and George (father) gave birth to their first child, a son named Alex. After he was born, being first time parents and having trouble with breastfeeding, Family B spend a lot of time at the pediatrician’s office and Alex was always on the side of slightly being failure to thrive. Debbie stated, “The pediatrician would always say, ‘Okay, let’s wait and see if he gains at least 5oz in the next two days before we do testing.’ But he would gain exactly 5oz, and that went on for about two weeks. We would have to weigh him before and after breastfeeding, too.”

At about two weeks of life, Alex’s doctor suggested that Alex be tested for CF. Debbie’s initial response was “it can’t be CF, I tested negative for the CF mutation.” Then she decided to go ahead with the testing because the Alex’s doctor said that Alex might have a rare CF mutation that would not have been included in the panel of testing Debbie had done. Results of Alex’s testing revealed that he had the most common CF mutation. This prompted Debbie to check back with her health care provider about her own testing. It turns out that when Debbie was pregnant she had agreed to undergo carrier screening for CF. However, a mistake was made and although the test was ordered and the blood was drawn, the test was never run. Debbie never received any notification about her test results because they had told her they would “call if it was abnormal.”
Because of this error in how her carrier screening for CF was done, Alex’s diagnosis came as a total shock. When discussing this, Debbie stated, “Yeah, if the testing hadn’t gotten messed up and we would have known. We wouldn’t have moved away from Baltimore. We had family and friends in the area and we had great insurance. Here, the insurance process has been a nightmare.”

In present day, Debbie and George have their son, Alex, who is thirteen, and a daughter, Gabby, who is eight. Debbie works full time as a Nurse Practitioner and George works full time as a counselor. For the first few years of Alex’s life, Debbie worked nights and George worked days. Although that is not the case anymore, they had to figure out the logistics of having full time jobs and taking care of two children, one of which has CF. Debbie and George do not have any family that lives in the area so when Alex is hospitalized, it is hard on their family, especially Gabby. Debbie and George have to flip flop from the hospital and home to take care of each child. Gabby is very sensitive and will state, “I just wish my family could be together.” Family B has insurance through Debbie’s job, but because insurance is so vital to taking care of Alex, she feels as if she can’t leave her current job. They have also reached the point of having conversations with Alex about longevity of life and that has been a difficult topic for Alex to handle. The treatments and care Alex goes through continue to be very time consuming. A stressor that has become more difficult to manage now that he is a teenager is absence from school for appointments and hospitalizations. He has more assignments and tests to make up on his own. George elaborated on this by stating, “Now it is more of a problem that over a 2-3 week period he might have six or so appointments so he is missing half-a-day multiple times in a few weeks and when he isn’t responsible and doesn’t follow-up, he will fall behind a little bit.”
Family Appraisal

Debbie and George seem to have adapted well to Alex’s diagnosis fairly quickly. With Alex being their first child, their life as a family was all about Alex and his CF during the first few years of his life. They reported that they couldn’t imagine their life any different than it was. Once their daughter was born, they felt as though their life became normalized. Debbie explained that being a big brother has been life-changing for Alex. Alex and Gabby have formed a very special relationship. Debbie stated, “There is not a regret in my mind. To see him as a big brother has been fabulous and for him to have that experience... and she has issues all on her own with her girl drama but the stresses have seemed to diffuse.” Both parents view their situation as something they have handled and can handle together, no matter what happens. Gabby has said that she wants to become a doctor so she can cure CF. They view Alex’s CF as requiring a team effort to conquer. They know he needs to be active and Debbie stated, “He has always been active but part of that is us. We are an active family and active people in general but we want him to keep up his lung function.”

Family Resources

The existing resources that Family B has includes: Debbie’s nurse practitioner career, strong marriage, good insurance, and access to Duke and UNC’s Cystic Fibrosis clinics. Debbie also started a night out for CF moms and they are a part of UNC’s advisory board. They are both involved in telling their story and advocating for families with CF. Debbie is involved in the community with CF moms and receives support from them.

Although Family B does not have any family close by, they have several friends in the community who can help them out with logistical problems. Family B also has access to therapists who have helped Alex and his family walk through stressful times in life. They have
also been very lucky to have schools that have been accepting and accommodating of Alex’s needs.

**Problem Solving and Coping Skills**

Both Debbie and George have great problem solving and coping skills. They agreed that they are good at figuring out problems and coming up with different solutions for the same problem and figuring out which one works best. George stated. “I think we are really good at coming up with solutions and compromising when our solutions aren’t the same. We don’t always agree, but that is okay. We can always end up resolving whatever we are trying to do.”

Even though Debbie has the medical background, George is also very involved with Alex’s care. George will do every aspect of his care other than accessing his port. As seen in the interview, Debbie and George act as a united front in the care and discipline of both of their children and they are united in that fact that they both want the best for their children. Family B copes together as a family unit and are very open with their communication. Having such an open dialogue helps all of them work through the stressors of living with CF. Finding a therapist that can connect with Alex has also helped him overcome some difficult situations. For Debbie, being so involved with the moms in their CF group has also been a good resource for coping and for keeping up with the changes in the life of CF. Debbie and George also sought out genetic counseling on their own when they were trying to decide if they were going to have another child.

**Family Adaptation**

Family B has overcome many stressors in the last decade and they have done it without having family in the area. They have worked through the critical event of Alex’s diagnosis and the subsequent stressors together and have created positive outcomes. They have continued to
Family Experience CF

have open communication throughout. Alex was Family B’s first child and they quickly adapted to their new normal when he was diagnosed. When they had their second child, they didn’t see having another child as a stressor but as a welcomed relief that normalized and diffused a lot of other stressors in their life. They have continued to grow stronger as a family unit and have impacted the lives of others, especially other families with CF.

Discussion

Family A and B have both adapted to having a child with CF. Although they have dealt with some of the same stressors, they each have their own unique story. A major difference between Family A and Family B is that in Family A, the youngest child has CF and he is a twin, while in Family B, the oldest child has CF. These differences create different family dynamics. Family A has an older brother who took on the role of caretaker in some ways and wants to teach other people about CF. It also has made Dawson more compassionate for other people. Family B has a younger sister who adores her older brother and looks up to him. When the family separates, it is hard on her and has helped her to become a sensitive person.

For both families, a major stressor in their pileup of demands was the significant amount of time it takes to do the CF treatments. However, the treatment demands are somewhat different in the two families. Because of this, they each have their own set of obstacles to overcome. In Family A, because Walker is six, Jan still does the majority of care and that is difficult when she also has two other children who have their own needs. In Family B, Alex, who is thirteen, can do a lot of his own care, but he at times lacks the necessary motivation and needs to be reminded to do his treatments and take his enzymes. While, Debbie and George do not have to physically help as much with his treatments, they still need to be heavily involved to make sure the treatments get done. Both families explained that the preparation and cleaning of for treatment
takes an overwhelming amount of time. It is also important to note that in Family A, Richard works a full time and Jan is able to stay home, while in Family B, both Debbie and George work full-time jobs.

While both Family A and B had a strong marriage with open communication, they have different external resources. Family A relies heavily on their faith and has family close by to help when needed. Family B does not have family able to help but they function very well as a family of four and are able to maintain normalcy in their routine. Both families have utilized therapists to help their child with CF overcome certain milestones and stressors. Family A and B both believed their child has a purpose and their family has become stronger because of living with CF. Although Family A’s younger child has CF and Family’s B older child has CF and they each had distinct challenges and decisions to make, they each looked at their situations as a challenge to overcome and were able to adapt while maintaining family stability.

This study examined how families adapt to living with a child with CF. Previous research focuses mainly on the individual who has CF and not the family as a whole. The results of this study provide a basis for understanding the family experience. By revealing some of the common stressors and resources, as well as family specific stressors and resources, the results have implications for nurses. The ability of nurses to identify particular stressors for families with CF will enable them to provide better care for these families and it will allow them to better educate other families who are experiencing similar stressors. Further research on the family experience of living with CF will increase our knowledge and provide more resources for CF families.

Although this study is a good basis for understanding how families live with CF, there are limitations. The biggest limitation of this study was the small sample size. A larger sample of families from diverse backgrounds with children of different ages would make the results more
generalizable. Further studies need to be done to assess the specific stressors families living with CF go through and to have a deeper understanding of how these families adapt. Findings from this and future studies will help nurses more effectively assess, educate, and guide families affected by CF. This in turn will help families maintain a healthy family unit as they face the ongoing challenges associated with living with CF.

**Implications for Practice**

The results of this study provide a basis for understanding the family experience of living with CF from the viewpoint of having a young school age child and an adolescent. This study shows that the pileup of demands can remain high no matter the age of the child, but the type of demands can shift as the age of the child changes. In Family A, when caring for a young school age child that has CF, the responsibility of not only the treatments but also activities of daily living largely remain with the parents. In Family B, the child with CF is an adolescent and can perform his own treatments but the parents spend a lot of time reminding him to perform the treatments and following up to make sure they are completed. Both families had a pileup of demands but they were different. It is important for the nurse to know and understand what the demands of the family are and to know what role each family member takes in performing CF treatments so the nurse can adequately and effectively educate the right people and offer the sufficient support and resources to the right members of the family.

This study showed that even though Family A and Family B reached adaptation in different ways, they are both well adapted. Communication and problem solving skills were important in both families but again, these processes were different in each family. For the nurse to provide quality care for CF families, it is important to know how they communicate and problem solve. In Family A, Jan and Richard made decisions together and came up with
solutions together, but the solutions and care were mainly implemented by Jan. It would still be important to communicate updates and information to Richard but new information about updates would affect Jan more because she is the main caregiver when it comes to CF treatments. In Family B, Debbie and George both work full time and have equal parts in the care and treatment of Alex’s CF. They have open communication and problem solve together, but because they both are equally involved in the CF treatments and both implement solutions, it is important for the nurse to know that both of the parents need to be updated and understand changes in treatment. If the nurse understands the family as a unit and understands the pileup of demands, how they communicate, how they problem solve, and who actively participates in the treatments, better quality of care can be given to the patient and to the family.
References


Appendix A

**Interview Guide for CF**

"Tell me about how you first become aware that your child has Cystic Fibrosis?"
- Who first talked with you about the diagnosis?
- "Who was with you when you were informed of your child’s diagnosis."
- "What was it like for you and other family members to hear the diagnosis?"
- "Who do you think had the most difficult time?"
- "Is there anything you would have liked to have had done differently."

"What kind of help and support did you find the most helpful initially?"

"Tell me about what it was like for you and your family the first year after your child with Cystic Fibrosis was born."
- "What kinds of things were most helpful to you initially?"
- "If there were things that made it more difficult, what were they?"

Tell me about what it was like for you and your family in the years between that initial year and now/
- "What kinds of things were most helpful to you?"
- "If there were things that made it more difficult, what were they?"

"How do you think you and your family are doing now.?"
- "What are the major challenges you are currently facing?"
- "What kinds of things help you deal with these challenges?"
- "What kinds of changes have you made in your life since your child with Cystic Fibrosis was born?"

If you had to write a book about your family’s experience of raising a child with Cystic Fibrosis, what would the title of that book be?
  - How about the chapters?
  - What would the main message of your book be?
  - What advice do you have for health care providers who care for children with Cystic Fibrosis and their families?