Influence of Contextual Factors and Self Efficacy on Self-Management in Parents of Children with Cystic Fibrosis

Erin B. Booth
Virginia Commonwealth University

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INFLUENCE OF CONTEXTUAL FACTORS AND SELF EFFICACY ON SELF MANAGEMENT IN PARENTS OF CHILDREN WITH CYSTIC FIBROSIS

A dissertation submitted in partial fulfillment of the requirements for the degree of Doctor of Nursing at Virginia Commonwealth University

Erin Bishop Booth
East Carolina University, B.S.N
Virginia Commonwealth University, M.S.

Director: Jeanne Salyer, Ph.D., RN
Associate Professor, Adult Health Nursing
School of Nursing

Virginia Commonwealth University
Richmond, Virginia
December, 2017
ACKNOWLEDGMENTS

I would like to express my appreciation to a number of individuals who have helped me with this project. First and foremost my academic advisor/dissertation chair, Dr. Jeanne Salyer. Dr. Salyer has been a source of persistent encouragement and guidance throughout coursework and the dissertation process. Also a special thanks to my other committee members Dr. Suzanne Ameringer, who stepped up a served as my advisor during coursework and lead me though my pilot study work. Dr. Edmond Wickham who helped me with survey development for my first pilot study and provided contributions towards the proposal. Dr. Kathleen Sawin who was critical in helping identify the theoretical framework used in my project. I would also like to acknowledge Dr. Lauren Goodloe, although no longer with us, who encouraged me to start this journey many years ago.

I also want to extend acknowledgment to the clinic staff at the Children’s Hospital of Richmond’s pulmonary (Cystic Fibrosis) clinic for being so welcoming and helpful during data collection. Also I would like to extend appreciation to the participants of this study who willing completed the surveys in hopes to help other families in the future.

I would also like to thank Keith Sledge (and family), founder of Pharmaceutical Specialties Inc. for giving me the opportunity to work and further my studies with both a flexible schedule and tuition assistance.

Finally, a special thank you to my supportive husband Jordan, and my children, Jackson and Ford. You have all been patient during this long process. I could not have done it without you. This would not have been possible without all of the gentle nudges and the support from all of my other family and friends.
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INFLUENCE OF CONCEPTUAL FACTORS AND SELF-EFFICACY ON SELF-MANAGEMENT IN PARENTS OF CHILDREN WITH CYSTIC FIBROSIS

A dissertation submitted in partial fulfillment of the requirement for the degree of Doctor of Philosophy at Virginia Commonwealth University.

Virginia Commonwealth University, 2017

Director: Jeanne Salyer, Ph.D., RN, Associate Professor
School of Nursing, Virginia Commonwealth University

Cystic Fibrosis, a life threatening autosomal recessive genetic disease, is characterized by a defective gene resulting in the production of thick mucus that obstructs the lungs and pancreas. CF requires intensive management performed at the home. An initial pilot study was performed to describe knowledge of CF related diabetes (CFRD) in adults with CF. The findings of this study, which demonstrated that adults with CF lacked sufficient knowledge about CFRD confirmed the need to explore additional self-management requirements guided by a theoretical framework. The second study presented in this dissertation used the Individual and Family Self-Management Theory (IFSMT) to describe context (condition-specific and individual and family factors) and process (self-efficacy and knowledge) and outcome (family self-management)
variables for caregivers of children with CF. It also compared differences in context, process, and outcomes in caregivers based on socioeconomic status (Medicaid vs. private insurance), and explored correlations among context, process, and outcomes. Participants for this cross-sectional descriptive study were caregivers of children with CF who had been diagnosed with CF for at least 9 months. Participants completed a demographic survey and questionnaires that included measures of perceived disease severity (VAS), depressive symptoms (Patient Health Questionnaire), self-efficacy (Perceived Health Competence Scale, Mountain West Cystic Fibrosis Consortium Questionnaire), knowledge (CF Knowledge and Attitudes Questionnaire), and self management behaviors (Self-Management Behaviors Questionnaire). Additional information was retrieved from the child’s medical record and included demographic information as well as height/weight/BMI, pulmonary function test results, medication profile, and insurance status.

Participants were primarily female caregivers with high self-efficacy, and average knowledge. The children with CF had moderate treatment complexity and normal/mild impairment in lung function. Deficits were noted in caregivers’ reproductive and genetic knowledge. This study found differences between Medicaid and private insurance groups related to knowledge. There were significant correlations between disease severity and CF specific self-efficacy and nutritional surveillance as well as general self-efficacy and respiratory surveillance.

These findings confirmed that the IFMST would provide a consistent framework to guide future studies aimed at identifying factors that influence self-management behaviors of CF in patients and their caregivers.
Chapter 1

STATEMENT OF THE PROBLEM

Background and Significance

Cystic fibrosis (CF), with a prevalence of 1 in 3500 live births, is the most common life-threatening autosomal recessive genetic disease in the United States. Cystic Fibrosis is characterized by a defective gene that results in the production of thick, tenacious mucus that obstructs the lungs and the pancreas. CF is a multisystem disease that primarily affects the respiratory and gastrointestinal systems (Table 1). Co-morbidities are common among patients with CF and include gastrointestinal symptoms, nutritional failure, diabetes, bone disorders, sinus problems, and mental disorders. (Fields, Michel, Butler, Kriss, & Albers, 2006; Moran et al., 2009; Sawicki, Sellers, & Robinson, 2008).

In the 1950’s, a diagnosis of CF resulted in a life expectancy of only 15 years; however, with continued advances in medical and nutritional management, the life expectancy has improved significantly and is now 43 years. Of the approximately 30,000 children and adults in the United States with CF, more than 50% are over the age of 18 years (Registry Highlights, CFF, 2016). The improved life expectancy has been achieved by employing complex management including multiple interventions to both prevent and treat manifestations of CF and its co-morbidities.

Increasing survival rates in CF are, in large part, due to more aggressive management of the disease. The regimen may include nebulized or oral antibiotics, enzymes or nutritional supplements, insulin, chest physical therapy and exercise. The majority of CF patients require care at home. Regardless of the age of the individual with CF, management is rarely a solo task;
Table 1: Manifestations related to Cystic Fibrosis

<table>
<thead>
<tr>
<th>Organ</th>
<th>Manifestation</th>
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<tbody>
<tr>
<td>Respiratory</td>
<td>Decreased mucociliary clearance, viscous mucus, airway inflammation, abnormal mucus traps, bronchiectasis</td>
</tr>
<tr>
<td>Nasal Manifestations</td>
<td>Chronic sinus infections, viscous mucus, polyps</td>
</tr>
<tr>
<td>Pancreatic</td>
<td>Recurrent pancreatitis, obstructed pancreatic ducts, malabsorption of vitamins A, D, E, K</td>
</tr>
<tr>
<td>Bone</td>
<td>Osteoporosis, low bone mineral density, arthritis</td>
</tr>
<tr>
<td>GI</td>
<td>Steatorrhea, meconium ileus, chronic elevation of liver enzymes, cirrhosis, portal hypertension, delayed gastric emptying</td>
</tr>
<tr>
<td>Reproductive</td>
<td>Men: azoospermia/infertility Women: reduced fertility</td>
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family members including parents, grandparents, siblings, and spouses often play an active role in assisting with care.

Chronic Illness

Chronic illnesses are typically defined as illnesses with a lengthy duration (longer than 3 months) that usually develop slowly. Common childhood chronic illnesses include asthma, diabetes, and cystic fibrosis. Childhood chronic illnesses present unique challenges to families who have to navigate a complicated treatment regimen while balancing other family responsibilities.

Many chronic diseases, including CF, have significant implications for the psychosocial well-being of the family (Barlow & Ellard, 2006). Furthermore, the need to manage chronic
conditions is increasingly recognized as the responsibility of the individual and family (Ryan & Sawin, 2009). For example, chronic pediatric respiratory diseases such as CF and asthma require patients and families to engage in ongoing monitoring and treatment to maintain their health.

*Self-Management*

Self-management has been defined in various ways in the literature. Richard and Shea define self-management as the ability of the individual, in conjunction with family, community, and health care professionals, to manage symptoms; treatments; lifestyle changes, and psychosocial, cultural, and spiritual consequences of health conditions (Richard & Shea, 2011). Self-management requires several general and disease specific skills. Examples include making decisions, partnering with health care providers, solving problems, managing the impact of the illness on various aspects of life, and taking action (Lorig & Holman, 2003). Self-management has been widely studied in chronic diseases such as diabetes, asthma, and heart disease. Self-management is especially important as it relates to chronic disease, where the patient is expected to be responsible for the day to day care over the length of the illness (Lorig & Holman, 2003). In CF, the ability to self-management is a lifetime task.

The management of CF is more complex than merely compliance with prescribed therapies; it requires patients and families to monitor symptoms and to modify therapies appropriately. Therefore, skills related to monitoring, decision-making, communicating, and coping are necessary to enable patients and families to perform self-management behaviors (Parcel et al., 1994). Parcel et al. postulated a model relating education variables and medical and interpersonal self-management variables to measures of health status. They found that knowledge and self-efficacy are important educational variables to address to enable caretakers of CF patients to be more effective in performing self-management behaviors. Previous research
has demonstrated that individuals with CF are able to perform 30-70% of these self-management tasks. Patients with CF, however, have more difficulty performing some treatments than others with the highest percent of self-management behaviors in antibiotics management and the lowest in physiotherapy behaviors (Abbott, Dodd, & Webb, 1996). Improved survival is in large part due to adoption of aggressive care guidelines and more frequent monitoring of complications. This has also lead to an overall increase in the complexity of treatment. With the number of available effective therapies increasing, patients and families may begin to make choices between the perceived cost or burden of particular self-management treatment behaviors and the perceived benefit. It will become increasingly important for caregivers and individuals with CF to understand the actual benefit of treatment in order to make informed choices.

Because CF is a multi system disease, the treatment regimen is multi faceted. As with any treatment regimen there are both barriers and facilitators. A study by George defined barriers to self-management as anything identified by the subjects as decreasing the likelihood they would follow a prescribed treatment regimen. The most common barrier was treatment burden (length, frequency, complexity of treatments). Additional barriers included social and work demands, forgetting, and absence of perceived health benefits, fatigue, and stigmas/embarrassment. This study also identified facilitators of CF self-management, Facilitators of self management are defined as patient perception of things to increase likelihood they would follow prescribed regimen. Several facilitators were identified. The most prevalent facilitator identified by the participants was the role of the CF Clinic. Participants acknowledged attending CF clinic, receiving feedback on pulmonary function test (PFT), and interacting with CF team promoted self-management. Other facilitators identified were support and reminders, presence of perceived health benefits, ease of completion, habit/routine, and distraction and rewards.
Knowledge

The role of accurate disease knowledge as it relates to self-management cannot be overlooked. Not only is it an important predictor of treatment adherence but it can also impact patient “buy in” by making the patient more informed and involved in their treatment decisions. (Balfour et al., 2014; Martin, Williams, Haskard, & Dimatteo, 2005). There are few studies that address understanding patients or parent’s knowledge of CF, and those that exist address specific issues (i.e. reproductive, nutrition). A higher level of knowledge has been correlated with better disease management.(Balfour et al., 2014; Parcel et al., 1994). Because the crux of CF care hinges on self management and self management hinges on knowledge, it is critical to have a strong knowledge base. Siklosi el al developed, validated, and implemented a disease knowledge questionnaire for adult patients with CF. They found that although adult CF patients understood most aspects of their disease, knowledge deficits did exist. The deficits were primarily related to genetics and reproduction. They found no signification associations between CF knowledge scores and socioeconomic factors but did find signification associations between scores and gender and education attainment. Being female and having higher education attainment were both independent predictors of higher CF knowledge scores. Balfour et al. indentified knowledge gaps including misinformation on the use of antibiotics relating to resistance, and understanding the mechanisms behind airway obstruction. They also identified a gap in knowledge around how and why airway clearance therapies work.

A 2003 study by McCabe highlighted gaps in dietary knowledge and enzyme application (McCabe, 1996). Studies have also shown that both males and females lack disease specific reproductive health knowledge and further identified the is a need for specific sexual and reproductive education (Gage, 2012). A recent study by Dashiff et al (2013) looked at a group of
adolescents (15-19 years old) who had been diagnosed with CFRD for at least a year. This descriptive study aimed to answer questions regarding parents’ and adolescents understanding of CFRD and its management and parents’ and adolescents’ perceptions of adolescents’ self-management of CF and CFRD. Although there was variation in level of understanding of CFRD from participant to participant, overall the understanding was rudimentary and inaccurate (Dashiff, Suzuki-Crumly, Kracke, Britton, & Moreland, 2013). The results of this study suggest that parent and adolescent education about CFRD needs to be improved and reinforced. Results of this study coupled with the current literature suggest that knowledge gaps exist in the both the adult CF as well as in caregivers to children with CF.

*Self-efficacy*

Self-efficacy beliefs are referred to as one’s beliefs in his or her abilities to perform a particular behavior effectively and successfully. The concept of SE originated from the social cognitive theory (Bandura 1989) which suggests that self-efficacy beliefs are largely derived through one’s personal accomplishment history in a given task. Research on the self-management of other chronic illnesses (diabetes, COPD, asthma) has demonstrated that self-efficacy is a critical determinant of learning and performance of self-management behaviors. Thus, self efficacy is a critical factor for successfully managing the challenges of a chronic condition such as CF.

**Theoretical Framework**

There is limited literature spanning more than 20 years addressing topics such as self management, knowledge, and self efficacy in the CF population, however this research has either been atheoretical or was guided by a variety of conceptual/theoretical frameworks making it difficult to make meaningful connections among variables important for chronic disease
management. The middle-range theory of Individual and Family Self-Management (IFSMT) (Figure 1) provides a theoretical framework to guide research on self or family management of CF because it suggests that self-management (SM) is a complex dynamic phenomenon consisting of 3 dimensions: context, process, and outcomes (Ryan & Sawin, 2009). Individual and family SM includes incorporation of health-related behaviors into an individual or family’s daily functioning. This theory combines and expands on previous work related to individual SM and adds a focus on dyads within the family or family unit. Although there are established relationships among variables in the model, a descriptive theory reveals the substance of a situation however does not provide structured links showing specific relationships among components. When individual SM is viewed as a systems theory, a change in one component of the system leads to changes in the entire system.
Figure 1: Individual and Family Self Management Theory
Combining the individual and family perspectives improves our understanding of shifts in balance and roles as family members often evolve and change. For individuals and families, the management of chronic health conditions leads to improving health outcomes, enhanced quality of life and realignment of healthcare expenditures. (Ryan & Sawin, 2009).

Statement of Purpose

The purposes of this study were to: (1) describe condition-specific factors (severity of child’s condition, complexity of the child’s treatment) and the social environment (SES, depression) for caregivers of children with CF, (2) describe caregiver self-efficacy, knowledge, and family self-management behaviors in caregivers of children with CF, (3) compare differences in context (depression), process (knowledge, self-efficacy) and outcomes (self-management behaviors) in caregivers based on SES (Medicaid vs. private insurance), and (4) explore correlations among context, process, and outcomes.

Summary

Guided by the Individual and Family Self-Management Theory (Ryan & Sawin, 2009), the purpose of this study is to examine the effects of context (condition specific factors and physical and social environment) on outcome (self-management). There are currently no studies using the IFSMT in CF.

In Chapter 2, we will review a pilot study that explored knowledge, specifically related to CF related diabetes, in adults with CF. This study helped form the background for the subsequent study (presented in Chapter 4) by recognizing the need for a theoretical framework, as well as the need to explore factors in additional to knowledge.
A SURVEY OF KNOWLEDGE RELATED TO CYSTIC FIBROSIS RELATED DIABETES

Abstract

Objective: Cystic fibrosis (CF) is the most common life threatening autosomal recessive genetic disease in the United States. Cystic fibrosis related diabetes (CFRD) is the most common co-morbidity of CF, as individuals with CF age, the likelihood of developing diabetes increases. CFRD leads to decreased lung function, poor nutritional status, and decreased survival rates. The extent to which individuals with CF know about CFRD is unknown.

Research Design and Methods: A cross sectional descriptive design was used. Adults (>18 yrs) with CF were recruited during an outpatient CF visit and asked to complete a 15-item CFRD Knowledge Survey. The survey included 10 items on knowledge of CFRD, 2 items on obtaining information about CF, and 3 items on experience with diabetes.

Results: Twenty-six individuals participated. Regarding knowledge, 92% had heard of CFRD, 65% knew symptoms of CFRD, and 58% knew how the diagnosis was made. A majority (65%) reported they seek information related to CF only when necessary. The total knowledge score was significantly correlated with the experience score ($r=0.50$, $P=0.009$), and understanding the importance of knowing that you have CFRD ($r=0.80$, $P<0.001$). The experience score was also significantly correlated with understanding importance of knowing if you have CFRD ($r=0.60$, $P=0.001$). Understanding the importance of knowing if you have CFRD was significantly correlated with being diagnosed ($r=0.43$, $P=0.028$).
Conclusion: As the number of individuals diagnosed with CFRD continues to increase, both pediatric and adult endocrine nurses need to be educated and equipped to successfully manage this patient population.
Cystic fibrosis (CF), with a prevalence of 1 in 3500 live births, is the most common life threatening autosomal recessive genetic disease in the United States. In the 1950’s, a diagnosis of CF resulted in a life expectancy of only 15 years; however, with advances in medical and nutritional management, life expectancy, which is now close to 40 years, has improved significantly. Of the approximately 30,000 children and adults in the United States with CF, more than 50% are over the age of 18 years (1).

Improved life expectancy is a positive outcome; however, it has led to the need to address previously overlooked long-term complications and co-morbidities. Impaired reproductive health, bone disease, depression, glucose intolerance, and cystic fibrosis related diabetes (CFRD) have all become integral to successful management of the aging CF population. CFRD, the focus of this study, is common and patients are not well informed of the symptoms and effects of this serious co-morbidity.

CFRD is the most common co-morbidity occurring in CF patients, occurring in approximately 20% of adolescents and up to 50% of adults (2). However, prevalence may be underestimated due to differences in screening protocols as well as lack of routine screenings. CFRD is primarily a result of insulin deficiency with some element of insulin resistance. Initially, alterations in glucose tolerance in CFRD are seen in the postprandial period, followed by a progressive decline in insulin secretion, leading to impaired glucose tolerance (IGT) and ultimately CFRD. As yet, increased age and the presence of exocrine pancreatic deficiency are the only predictive indicators for developing glucose intolerance or CFRD (2). The impact of CFRD cannot be disregarded. Individuals with CFRD have higher rates of lower lung function (3), poor nutritional status (4), and decreased rates of survival (5) compared to individuals with CF but without CFRD.
Early diagnosis is critical because clinical deterioration begins 6-24 months prior to diagnosis. Therefore, it is important for health care providers to be aware of what their patients and caregivers know about CFRD. Moran et al (2) recommended that patients should be well informed on the frequency of CFRD and the benefits of treatment as well as be provided education and psychological support. To date, there is limited published information on what individuals’ with CF know about CFRD. In order to adequately address educational needs in this population we must first understand their current knowledge and awareness of CFRD.

The purpose of this pilot study was to evaluate the understanding of CFRD in adults with CF. Specifically, we aimed to a) describe knowledge and experience of CFRD in adults with CF, b) examine the relationship between knowledge of CFRD and previous experience or exposure to diabetes and c) describe the sources by which adults with CF obtain information related to their CF disease.

Methods
Participants
Participants were recruited through the pulmonary clinic at the Virginia Commonwealth University Medical Center. The clinic treats approximately 140 patients with CF and approximately 45% of the patients seen in the clinic are over 18 years old. The gender breakdown is 51% males, 49% females and ethnicity is similar to what is seen across the United States with 94% Caucasian and the remaining Hispanic or African American. Inclusion criteria were age 18 and older and a diagnosis of cystic fibrosis. Exclusion criteria were pregnancy and/or inability to read or write English. There were 28 participants approached for participation and all chose to participate.

Procedure
The Institutional Review Board approved the study. Recruitment occurred by either the healthcare provider or the researcher approaching patients at the time of their clinic visit to determine eligibility and interest in participation. If interested, an information sheet with details concerning the study was provided and served as the implied consent form. Interested individuals completed the survey.

Instrument

The CFRD Knowledge survey was initially developed by the researcher and formally evaluated for content and comprehensiveness by content experts including pulmonary and endocrinology clinicians. The survey consists of 15 items: 1 item assessing presence of CFRD, 8 items on knowledge of CFRD, 1 item to assess how important participants feel it is to know about a diagnosis of CFRD, 2 items on how often and where participants seek information on CF, and 3 items on previous experience with or exposure to individuals with diabetes.

A total score was calculated for knowledge; six of the eight items were used to calculate the score. To assess if participants were aware of the relationship between CFRD and lung function and weight, four questions were included, two were distracters (not used in the score) and two assessed knowledge of these relationships. For all knowledge questions, response options were either yes or no, with opportunities for participants to provide comments. The comments were not used in scoring however; the purpose was to glean additional information not specifically addressed in the questions. A score was generated by totaling the number of yes responses. The knowledge score could range from 0-6 with higher scores indicating more knowledge.

A total experience score was calculated from the 3 experience with diabetes items including experience with high blood sugar, experience with insulin, or a family member with
diabetes. Experience scores could range from 0 to 3, with a score of one or greater indicating experience.

Demographics and Disease Related Information

For sample description, participants were asked to provide age, gender, ethnicity, race, highest grade completed, employment status, marital status, how long they had been diagnosed with CF, and if they had ever been treated at another CF center.

Statistical Analysis

Demographic information was summarized with means (SD) or frequencies and percentiles. For aim 1, descriptive statistics were used to describe knowledge of CFRD. For aim 2 descriptive statistics were also used to describe the frequency and sources that adults with CF obtain information about their disease. For aim 3 Pearson correlations were calculated between total knowledge score and experience score, knowledge and experience scores and understanding the importance of knowing that you have CFRD. Analysis was performed using SPSS version 19.

Results

Sample Description

The mean age of the participants was 28.1 years (SD= 10.22; range =18-54). The majority were male (70%) and non-Hispanic or Latino (96%). Thirty-three percent had been previously diagnosed with CFRD. Table 1 includes additional sample characteristics.

Knowledge and Experience

The majority of participants (92%) had heard of CFRD, however only 62% knew they were at risk for developing it. When asked if they knew how the diagnosis of CFRD was made, 58% stated yes. Common responses when asked to explain how the diagnosis was made
included: blood sugar checks, glucose tolerance test, and Hemoglobin A1C. When asked if they knew the symptoms of CFRD, only 65% of participants responded yes. The symptoms that were most frequently identified were increased thirst, increased urination, weight loss or inability to gain weight and dizziness.

The average knowledge score among all participants was 3.85 however, when broken into subgroups (diagnosed with CFRD and not being having CFRD) average knowledge score of the non CFRD group was 3.28 versus an average score of 5 for those that have CFRD.

Only 35% of participants stated that CFRD was related to lung function and 69% acknowledged the relationship to weight. When asked how important it was to know if they have CFRD, most (86%) participants felt it was either essential or very important. The majority of participants (70%) indicated previous experience with diabetes. The majority (70%) of participants had some experience or exposure to diabetes, and more than half (54%) had been told they had high blood sugar in the past. Thirty-eight percent of participants had previously been on insulin and 42% had members of their family with diabetes.

Knowledge was significantly correlated with experience ($r = 0.50, p = 0.009$), such that more knowledge about CFRD was associated with more experience. Knowledge was also significantly correlated with understanding the importance of knowing you have CFRD ($r=0.80, p <0.001$). Higher knowledge was related to a greater value being placed on understanding the importance of knowing you have CFRD. Experience was significantly correlated with understating the importance of knowing if you have CFRD ($r=0.60, p=0.001$). Understanding the importance of knowing if you have CFRD was significantly correlated with being diagnosed with CFRD ($r=0.43, p=0.028$).
Information

We were also interested in knowing how often, from whom, and where participants seek information about their disease. The majority (63%) reported they seek information when they need it, 19% seek information monthly, 11% seek weekly, and 7% stated they never seek information. Doctors were the most common source of information (77%), followed by internet (69%), and clinic (62%). Participants were able to list additional sources and the top two sources included two websites: the Cystic Fibrosis Foundation (www.cff.org) and Patients Like Me (www.patientslikeme.org).

Conclusion

As medical advancements and life expectancy have improved, many children with CF are living well into adulthood. Questions regarding age, long-term complication and co-morbidities have become more relevant but are not yet fully understood. CFRD represents a gap in disease-specific knowledge. This study represents the first attempt to understanding what patients know and understand about CFRD.

Knowledge and Experience

There is limited literature that addresses disease specific knowledge in the CF population. A 1996 study by McCabe (6) highlighted gaps in dietary knowledge and enzyme application. Studies have also shown that both males and females lack disease specific reproductive health knowledge and further identified the is a need for specific sexual and reproductive education (7-9). A recent study by Dashiff et al (10) looked at a group of adolescents (15-19 years old) who had been diagnosed with CFRD for at least a year. This descriptive study aimed to answer questions regarding parents’ and adolescents understanding of CFRD and its management and parents’ and adolescents’ perceptions of adolescents’ self-management of CF and CFRD. To
assess both the adolescents and parents’ understanding of CFRD, participants were asked, “if you were talking with a friend, what would you tell them CFRD is?” The findings demonstrated limited understanding of CFRD by both parents and adolescents ranging from inability to explain basics of CFRD, to inaccurate explanations of the disease often based on inappropriate generalizations to other types of diabetes. Although there was variation in level of understanding of CFRD from participant to participant, overall the understanding was rudimentary and inaccurate. The results of this study suggest that parent and adolescent education about CFRD needs to be improved and reinforced (10).

Siklosi el al (11) recognized the need to develop and validate a tool that would investigate general knowledge in the adult CF population. The results demonstrated that although adult CF patients understand most aspects of their disease, knowledge deficits did exist. The deficits were greater related to genetics and reproduction. They found no signification associations between CF knowledge scores and demographics, socioeconomic factors, clinical details but did find signification associations between scores and gender and education attainment. Being female and having higher education attainment were both independent predictors of higher CF knowledge scores.

This study adds to the currently literature highlighting a gap in CFRD knowledge. Since this study only included participants who had been diagnosed with CFRD, it can be assumed that patients and families who had not been diagnosed may have an even greater need for appropriate CFRD information related to screening, symptoms, and significance to aid in early recognition and perhaps earlier diagnosis.
Sources of Information

A concerning finding is that the majority of participants stated they only seek information when needed. In general, patients with CF believe they have a high level of disease-specific and treatment knowledge. Although this may be true, a problem exists if these individuals are not fully aware of co-morbidities and complications of the disease and, therefore, are not readily seeking information. Consistent with our findings, which identified doctors and the clinic as two of the most common sources of information, the reproductive literature has previously identified CF providers as the preferred source of information. In addition to providers, the internet was a popular method for obtaining health information. Patients Like Me is a site where patients can provide personal stories and health data with the goals of obtaining information about medications, supplements, and devices, and to connect easily with patients that have the same condition. CFF.org is the official website of the Cystic Fibrosis Foundation. This site provides a wide range of information for the public, patients, families, and clinicians. Information about CF, the foundation, living with CF, and research is included. This may reflect a the current trend of the internet’s growing popularity or may also be influenced by the restricted face-to-face contact patients with CF have with other patients with CF. This restriction limits the ability to participate in traditional support groups perhaps fueling the popularity of sites like Patients Like Me.

Limitations

There are several limitation of this study. Generalizability of results is limited due to a small sample size and convenience sample. Although CF occurs equally in males and females, there were a higher percentage of males in this study. The most plausible explanation for the difference would be that this was a convenience sample. Another limitation was the use of an
investigator developed survey tool. Further testing of this survey is needed with a larger sample size in a variety of settings. Nevertheless, despite these limitations, this study provides important insights that can be used to guide future studies.

Results of this study suggest that knowledge gaps exist in the adult CF population related to CFRD. Specifically, participants lack an understanding they are at risk, do not know the symptoms of CFRD and do not understand the relationship between CFRD and lung disease. These results, although not generalizable can guide future studies. As the CF population continues to age and life expectancy increases, it will become critical to understand knowledge gaps and provide appropriate and effective educational interventions. Studies are needed to explore patients, parents’ and caregiver’s knowledge and understanding of CFRD as well as the need to reinforce knowledge and understanding following diagnosis. Future studies will also need to examine the increased treatment burden associated with the additional diagnosis of CFRD as well as self-management support and guidance.
References

1. About CF [Internet] available from www.cff.org/AboutCF


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<th>Table 1: Sample Characteristics</th>
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<tr>
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**Sex**
- Male: 19 (70%)

**Ethnicity**
- Hispanic or Latino: 1 (4%)
- Non-Hispanic or Latino: 26 (96%)

**Race**
- Caucasian: 23 (85%)
- African American: 4 (15%)
- Other: 4 (15%)

**Level of Education**
- Grade 9-12: 1 (4%)
- Grade 12 or GED: 9 (33%)
- College 1-3 years: 12 (44%)
- College 4 years/graduate: 4 (15%)
- Graduate School/advanced degree: 1 (4%)

**Employment Status**
- Employed for wages: 9 (33%)
- Self employed: 1 (4%)
- Homemaker: 2 (7%)
- Student: 7 (26%)
- Unable to work: 5 (19%)
- Other (multiple status selected): 3 (11%)

**Marital Status**
- Married/Life partner: 9 (33%)
- Divorced/Separated: 1 (4%)
- Widowed: 1 (4%)
- Single, never married: 16 (60%)

**Treated at another CF Center**
- Yes: 13 (48%)
- No: 14 (52%)
Chapter 3
THEORETICAL FRAMEWORK AND METHODS

Theoretical Framework

Chapter 1 provided a general introduction to the middle-range theory of Individual and Family Self-Management (IFSMT) this chapter will delve further into the theory and highlight how the IFSMT would be used to guide research on self or family management of CF.

**Context.** Context is comprised of risk and protective factors. These factors are *Condition-Specific, Physical & Social Environment, and Individual and Family* factors that challenge or protect individuals and families as they engage in SM. Contextual factors influence the individual’s and family’s engagement in the SM process, as well as directly influence outcomes.

*Condition specific* factors can include physiological, structural, or functional characteristics of the condition that affect the amount, type, and nature of behaviors required to manage the condition. Condition specific factors also incorporate the complexity of the condition and/or treatment trajectory. *The physical and social environment* is comprised of factors such as access to healthcare, provider or setting transitions, transportation, neighborhoods, or culture that present barriers or improve ability for SM by the individual and family. *Individual and family factors* represent circumstances that enhance or diminish SM. Examples include cognitive status, perspectives, information processing, developmental stages, capabilities, literacy, or resourcefulness.
The treatment guidelines for CF recommend the use of a number of chronic maintenance medications, as well as airway clearance and regular exercise. This self-management regimen is time consuming and complex and requires proper use of devices. The complexity of treatment for individuals with CF is high, and estimates suggest that the self-management regimen takes two to three hours per day for routine care (Sawicki, Sellers, & Robinson, 2009). It has already been established that adherence to CF treatments is poor, typically around 50% (Eakin, Bilderback, Boyle, Mogayzel, & Riekert, 2011). A recent study by Sawicki et al. 2013 observed that although treatment complexity was highest in adults with CF, treatment complexity in all age groups increased over a 3 year period. In addition to the increase in treatment complexity being statistically significant, it is also clinically significant. Increases in treatment complexity, while necessary to improve health, pose challenges to self-management and adherence to therapies. In this case, high treatment burden and complexity would be a risk factor.

The physical and social environment is comprised of factors such as access to healthcare, provider or setting transitions, transportation, neighborhoods, or culture that can either prevent or enhance the ability to SM. Quitter et al. (2010) states that minority status and socioeconomic status (SES) contribute to variability in the clinical course of CF. (Quittner et al., 2010). They confirmed the findings of Schechter et al. and found that low SES was associated with worse pulmonary function, lower height and weight, and greater likelihood of Pseudomonas aeruginosa (Quittner et al., 2010). Possible explanations are due to increased family stress, exposure to environmental toxins, differences in disease self-management and culture perceptions of chronic illness. (O'Connor et al., 2002; Quittner et al., 2010; Schechter, Shelton, Margolis, & Fitzsimmons, 2001). Schechter et al. found that medically indigent patients with CF suffer more serious consequences of their disease than the general population with CF.
Medically indigent patients have a threefold risk for death, and survivors have significantly worse pulmonary function and growth. While decreased access to health care has been identified as an important cause of SES-related adverse outcomes in other populations, it does not appear to play an influential role in CF; as medically indigent CF patients were found to have the same number of outpatient specialty visits as privately insured cohorts. Possible alternative explanations to explain the disparity in health status in spite of similar medical appointments include nutritional inadequacies associated with poverty, a clear link exist between inadequate nutrition and more severe lung disease. Additionally patients with a lower SES status are more likely to be exposed to indoor and outdoor air pollution which can result in worsening lung disease. Other hypothesized causes are increased stress and its role in immune function and subsequently infection and decreased disease related knowledge.

**Process.** The key factors under Process are *Knowledge and Beliefs, Self-regulation Skills, and Social Facilitation.* IFSMT proposes that persons will be more likely to engage in recommended health behaviors if they have accurate information and accept health beliefs consistent with behavior. *Knowledge and beliefs* reflect information and perceptions about a condition or behavior and influence self-efficacy, outcome expectations, and goal congruence. Self-efficacy refers to the degree of confidence one has of his/her ability to successfully engage in a behavior under a variety of circumstances. Outcome expectations are a belief that engagement in a certain behavior will result in desired outcomes. Goal congruence is a person’s ability to resolve the confusing from competing or contradictory demands associated with health goals. *Self-regulation* is an iterative process that people engage in to achieve a change in health behaviors. This can include but is not limited to goal setting, self-monitoring, decision-making, planning, self-evaluation, and management of responses. *Social Facilitation* occurs when
relationships enhance an individual’s capacity to change. This can include social influence, support, and collaboration.

Outcomes. The primary outcomes are related to individuals and families; however, improvement of individual and family outcomes translates to improved outcomes for healthcare and systems. Outcomes include proximal (short term) outcomes that lead to attainment of distal outcomes. Proximal outcomes include individual and family self-management behaviors and Cost of Healthcare Services. Engagement in SM behaviors may or may not affect cost of health care services. Distal outcomes are defined as health status (as an indicator of disease trajectory), quality of life, and cost of health (indirect and direct costs).

Conceptual Framework

Guided by the Individual and Family Self-Management Theory (Ryan & Sawin), the purpose of this study is to examine the effects of context (Condition Specific Factors and Physical and Social Environment) on Outcome (self-management) in CF. (Figure 2)

Methodology

This section presents the design, sample, setting, and data collection procedures are well as the instruments and their psychometric properties, and data analysis plan.

Research Design

This study was conducted using a cross sectional descriptive design to describe condition-specific factors (severity of child’s condition, complexity of the child’s treatment) and the social environment (SES, depressive symptoms) for caregivers of children with CF, (2) describe caregiver self-efficacy, knowledge, and family self-management behaviors in caregivers of children with CF, (3) compare differences in context (depression), process (knowledge, self-
efficacy) and outcomes (self-management behaviors) in caregivers based on SES (Medicaid vs. private insurance), and (4) explore correlations among context, process, and outcomes.

Sample Selection and Setting

Permission was obtained from the Intuitional Review Board (IRB). A convenience sample of caregivers of individuals with CF was recruited from the pediatric pulmonary clinic at the Children’s Hospital of Richmond. Any caregiver, who was over the age of 18 and identified as a caregiver to a child/adolescent under the age of 18 with a diagnosis CF for at least 9 months was invited to participate. Caregivers were excluded if they were unable to read and write in English.

Instruments and Measures

The instruments that were used in this study included a demographic questionnaire, medical record review. In addition to the demographic questionnaire and medical record review other context variables were measured using a treatment complexity score, a visual analog scale (VAS), and the Patient Health Questionnaire (PHQ-9). Additional process variables were measured and included the Perceived Health Competence Scale (PHCS), the Mountain West CF Consortium, and the CF Knowledge and Attitudes test. The outcome variable was measured by the Self Management Questionnaire.
Figure 2: Individual & Family Self-Management Theory Applied to Family Self-Management of Children with Cystic Fibrosis
Individual Characteristics

Demographic questionnaire (Caregiver): A 14-item self report questionnaire provided information on the caregiver and included age, gender, race, marital status, highest education completed, employment status, relationship to the child with CF, and information regarding the number of other children in the family (see Appendix A).

Demographic characteristics (Child): The following characteristics of the child were collected as part of the caregiver questionnaire; gender, age and insurance status. The clinical characteristics of the child included; lung function, height/weight/BMI percentile, and medications/treatments were obtained from the medical record.

Context Variables

Treatment complexity. Treatment complexity can include the number of drugs, route of administration, dosing frequency, and additional directions for taking the drug for example, relationship to timing with foods, set up of device to deliver drug (George, Phun, Bailey, Kong, & Stewart, 2004). The Treatment Complexity Scale (TCS) was developed by Sawicki et al. (2010) in an effort to evaluate CF treatment complexity (see Appendix B). The TCS is a total score of 0-63, determined by assigning a complexity of 1, 2, or 3 for each therapy. The complexity score is based on daily frequency, duration, and ease of administration. The tool includes 37 chronic therapies that were captured as part of the longitudinal Epidemiology Study of Cystic Fibrosis (ESCF). Oral medications and metered-dose inhalers are assigned a score of 1 (20 items); once-daily nebulized therapies and pancreatic enzyme are assigned a score of 2 (8 items), and airway clearance, twice-daily nebulized therapies, use of oxygen, continuous positive airway pressure (CPAP), bi-level positive airway pressure (BiPAP), and insulin or enteral supplements are assigned a score of 3 (9 items). A total score is calculated by summing the
scores of all therapies and can range from 0-63. Higher scores indicate higher treatment complexity. Since the development of the tool, several new treatments for CF have been approved. The first is an oral medication that is taken twice a day. Similar to other twice a day medications this will be scored as 2. The second is a nebulized therapy, however, unlike other nebulized therapies which are administered once or twice daily, aztreonam is used 3 times a day. Although the treatment time is shorter (3-5 min compared to 10-20) it does require preparation and cleaning of nebulizer and will therefore receive a score of 3. With the additions of these two treatments the maximum score will be 68.

Disease severity. Disease severity was measured using information from pulmonary function testing. The pulmonary function testing was performed by the respiratory therapist as part of the routine clinic visit using the MCG diagnostics pulmonary function equipment with Breeze Suite © software (St. Paul MN). The forced expiratory volume in one second (FEV₁) was used and is classified into three groups: mild disease (FEV₁>70%), moderate disease (FEV₁ 41–69%) and severe disease (FEV₁<40%). This disease severity classification has been applied and adopted internationally in both the medical and psychosocial literature and is used as the standard for the epidemiological study in CF, and is the classification used in the Cystic Fibrosis Foundation (CFF) patient registry (Cystic Fibrosis Patient Registry, 2016 Annual Data Report). As an additional measure of disease severity, a visual analog scale (VAS) was used to gauge how the parent/caregiver viewed the child’s health (see Appendix C). Participants were asked to rate their perception of the child’s health by placing an X on a 10cm line. One side of the line (0 cm) represented death and the other end (10 cm) was perfect health. A score reflecting the position of the X on the line represented the perception of their child’s severity.
Depression. The Patient Health Questionnaire (PHQ-9) was used to screen for depression (see Appendix D). In 2015, the International Committee on Mental Health in Cystic Fibrosis began recommending the use of this tool. The PHQ-9 is brief, reliable, and valid with appropriate cut off scores for detecting depressive symptoms. The score ranges from 0-27 with a recommended interpretation as follows: 1-4 minimal depression, 5-9 mild depression, 10-14 moderate depression, 15-19 moderately severe depression, and 20-27 severe depression. A cut off score of ≥ 10 has a sensitivity and specificity of 88% for major depression. Internal consistence is high with Chronbach’s alpha 0.86-.89 (Kroenke et al., 2001).

Process Variables

Self-efficacy. To provide a broader assessment of self-efficacy, both general and disease-specific self-efficacy were considered. General self-efficacy was measured by the Perceived Health Competence Scale (PHCS) (see Appendix E). The PHCS contains 8 items to which responses are chosen from a 5 point Likert scale ranging from 1 = strongly disagree to 5 = strongly agree. There are 4 positively formulated items and 4 negatively formulated items. The four negatively worded items were re-coded so that a higher total score reflected self-efficacy (competence to manage their child’s health). The items add up to a total score with a range from 8-40; higher scores indicating higher levels of competence. The PHCS has been administered to wide variety of participants with a degree of health conditions. The psychometric properties suggest a high degree of internal consistency and satisfactory test-retest stability. Across five samples (young adult students, working adults, and persons with a chronic illness; rheumatoid arthritis) in the original study of the PHCS there was high internal consistency with Cronbach’s alpha ranging from 0.82-0.90 (Smith, Wallson, Smith, 1995).
The Mountain West Cystic Fibrosis Consortium Questionnaire (MWCFC-Q) developed by McDonald, Christensen, Lingard, & Walker, 2009 was used to measure caregivers disease-specific self-efficacy (see Appendix F). This Likert-type questionnaire (0=not at all confident to 10=completely confident) is divided into four sections 1) demographics, 2) food security, and 3) CF nutrition knowledge, and 4) confidence in CF management. In this study, only the 18 item section on confidence in CF management will be used. This section, which includes the three domains of CF care, allows the participants to rate their self-efficacy or confidence in each of these 3 domains: Task, Nutrition, CF Care center recommendations. Task measured the ability to manage CF care (pulmonary therapies and medications, nutrition addresses their ability to manage nutrition therapy and food related behaviors, and CF care center recommendations aimed to measure the participants perceived ability to follow the CF care center recommendations. The Cronbach alpha for the self-efficacy subscale has been 0.92 (McDonald et al, 2009).

Cystic fibrosis knowledge. CF knowledge was measured by the CF Knowledge and Attitudes Questionnaire developed by Siklosi, Gallagher and McKone (2010) (see Appendix G). The original questionnaire contains 49 fixed response questions, 2 open ended questions, and 9 questions combining both. The questionnaire is divided into three sections: demographics, CF knowledge test, and attitude and education questions. For the purpose of this study the CF knowledge test was used. The CF knowledge questionnaire is further divided into three sections: general knowledge (7 items), lung/GI issues (10 items), and reproductive/genetics knowledge (9 items). The CF knowledge test is scored by earning 1 point for each correct answer and 0 points for an incorrect answer or the choice “I’m not sure”. A maximum score of 32 is possible. The
Cronbach’s alpha for the overall test has been reported as 0.75. The interclass correlation coefficients ranged from 0.76-0.97 and indicate good test-retest reliability (Siklosi et al 2010).

Proximal Outcomes

*Family Self-Management of CF.* Self-management of CF was measured using The Self-Management Questionnaire for Cystic Fibrosis (SMQ-CF) (Sockrider et al., 1996). This instrument has both a caretaker and adolescent version, for this study the caretaker version was used (see Appendix H). The SMQ-CF measures behaviors that include skills characteristically used for management of both pulmonary and gastrointestinal symptoms in CF. The SMQ-CF is a 46 item written questionnaire using a 5 five point rating scale. Responses rank from 1 = never used to 5 = always used. Items address behaviors reflecting monitoring and treatment of respiratory infection, respiratory therapies, malabsorption and malnutrition. A ‘skip pattern’ is included for patients who do not have pancreatic insufficiency and also for those who have not had an experience with lower respiratory infections (bronchitis, bronchiolitis, or pneumonia). Due to the “skip pattern”, each of the sections (respiratory surveillance, respiratory treatment, CPT/RT, malabsorption, and nutritional surveillance) were examined separately based upon whether or not the treatment was required for the child. For example, all caregivers responded to the respiratory surveillance domain; however, if their child did not have an infection, the section on treatment could be “skipped”. Limitations in determining validity exist as there is no “gold standard” for comparison. Internal consistency was high with a Cronbach’s alpha coefficient of 0.95 for the total scale (Sockrider et al, 1996).

Data Collection

Participant recruitment was initiated following the IRB approval and a meeting with the clinic to review the project took place. Prior to the weekly CF clinic a pre-clinic meeting was
held and the list of patients was reviewed for participation. Participants were approached during the outpatient clinic visit by the researcher. They were informed about the study verbally and by review of the written consent, if they decided to participate informed consent (Appendix I) was obtained. Following the consent process participants were given a copy of the questionnaires. Participants had the opportunity to complete the questionnaires while at their visit or return them by mail via a pre-paid addressed envelope. Participants were counseled not to include any identifying information (i.e.: copy of consent) when returning by mail.

Research Subjects Protection

The only anticipated risk of participation in the study was the time it would take to complete the surveys. Participants were able to take breaks as needed. Participants names did not appear on the questionnaires and their answers were kept confidential. The questionnaires were kept in a locked file cabinet in the researcher’s office.

Statistical Analysis

All data were analyzed using SPSS (v. 24, Chicago, IL). Each instrument was evaluated for reliability using Cronbach’s alpha coefficients. Descriptive statistics were computed for sample demographics and all other variables using frequency distribution and percent for categorical variables and mean and SD based on distribution for continuous variables. Variables were inspected for normal distribution and means and SD were reported for those variables that were normally distributed. Medians values were provided for non-normally distributed values. Because the normal distribution assumption was not met, Mann-Whitney U test were conducted to compare insurance coverage groups (private, Medicaid) regarding depression, self-efficacy, knowledge, and self-management behaviors. Pearson’s correlation coefficient ($r$) was used to
determine relationships between context and process variables and self-management behaviors including treatment complexity, disease severity, knowledge, self-efficacy and depression.

Summary

The purpose of this study was to describe and explore the relationship between context, process and outcomes variables in caregivers of children with CF using a cross sectional design. The data was analyzed using SPSS (v.24). In Chapter 4 an additional description of the study as well as the results are presented in manuscript form.
Chapter 4

INFLUENCE OF CONTEXT AND PROCESS FACTORS ON FAMILY SELF MANAGEMENT IN CAREGIVERS OF CHILDREN WITH CYSTIC FIBROSIS

Abstract

**Purpose:** The purposes of this pilot study were to: (1) describe condition-specific factors and the individual and family factors for caregivers of children with Cystic Fibrosis (CF), (2) describe caregiver self-efficacy, knowledge, and family self-management behaviors in caregivers of children with CF, (3) compare differences in context (depression), process (knowledge, self-efficacy) and outcomes (family self-management behaviors) in caregivers based on SES (Medicaid vs. private insurance), and (4) explore correlations among context, process, and outcomes.

**Design and Methods:** Participants for this cross-sectional descriptive study were caregivers of individuals with CF who were under the age of 18 and diagnosed with CF for at least 9 months. The variables were selected using the Individual and Family Self-management Theory (IFMST) as a framework. Descriptive statistics were computed for sample demographics and all other variables. Mann-Whitney U test were conducted to compare insurance coverage groups (private, Medicaid) regarding knowledge, self-efficacy and depressive symptoms. Pearson’s correlation coefficient ($r$) was used to determine relationships between context and process variables and self-management behaviors including treatment complexity, disease severity, knowledge, self-efficacy, and depressive symptoms.

**Results:** Overall this study of primarily female caregivers was described as having high self-efficacy, and average knowledge. Deficits were noted in the areas of reproductive and genetic knowledge. This study found differences between Medicaid and private insurance groups related
to knowledge. There were significant relationships between perceived disease severity and CF specific self-efficacy and nutritional surveillance as well as general self-efficacy and respiratory surveillance. The children with CF in this study had moderate treatment complexity and normal to mild lung function impairment.

**Practice Implications** This was the first time that the IFMST was used to guide CF research. The IFMST would provide a consistent framework to guide future studies aimed at identifying factors that influence self-management behaviors of CF in patients and their caregivers. Based on the knowledge gaps identified, clinicians need to continually reinforce caregivers understanding of the disease and in particular with regards to reproductive and genetics knowledge. Additional studies are needed.

**Keywords:** Cystic Fibrosis, Treatment Complexity, Self-efficacy, Self-management, Individual and Family Self-Management Theory
Introduction

Cystic fibrosis (CF), with a prevalence of 1 in 3500 live births, is characterized by a defective gene that results in the production of thick, tenacious mucus that obstructs the lungs and the pancreas. CF is a multisystem disease that primarily affects the respiratory and gastrointestinal systems. Co-morbidities are common among patients with CF and include gastrointestinal symptoms, nutritional failure, diabetes, bone disorders, sinus problems, and mental disorders.

In the 1950’s, a diagnosis of CF resulted in a life expectancy of only 15 years; however, with continued advances in medical and nutritional management, life expectancy has improved significantly and is now 37.4 years. Of the approximately 30,000 children and adults in the United States with CF, more than 45% are over the age of 18 years ("About CF," 2017). The improved life expectancy has been achieved by employing complex management including multiple interventions to both prevent and treat manifestations of CF and its co-morbidities.

Increasing survival rates in CF are, in large part, due to more aggressive management of the disease. The regimen may include nebulized or oral antibiotics, enzymes or nutritional supplements, insulin, chest physical therapy and exercise. The majority of individuals with CF require care at home. Regardless of the age of the individual with CF, management is rarely a solo task; family members including parents, grandparents, siblings, and spouses often play an active role in assisting with care.

The middle-range theory of Individual and Family Self-Management (IFSMT) (Figure 1) provides a theoretical framework to guide research on self or family management of CF because it suggests that self-management (SM) is a complex dynamic phenomenon consisting of 3 dimensions: context, process, and outcomes (Ryan & Sawin, 2009). Individual and family self-
management includes incorporation of health-related behaviors into an individual or family’s daily functioning. This theory combines and expands on previous work related to individual self-management and adds a focus on dyads within the family or family unit. Although there are established relationships among variables in the model, a descriptive theory reveals the substance of a situation, however does not specify the strength and direction of relationships among components. When individual self-management is viewed as a systems theory, a change in one component of the system leads to changes in the entire system. Combining the individual and family perspectives improves our understanding of shifts in balance and roles as family members often evolve and change. For individuals and families, the management of chronic health conditions leads to improving health outcomes, enhanced quality of life and realignment of healthcare expenditures (Ryan & Sawin, 2009).

The purposes of this pilot study were to: (1) describe condition-specific factors (severity of child’s condition, complexity of the child’s treatment) and the social environment (SES, depressive symptoms) for caregivers of children with CF, (2) describe caregiver self-efficacy, knowledge, and family self-management behaviors in caregivers of children with CF, (3) compare differences in context (depressive symptoms), process (knowledge, self-efficacy) and outcomes (self-management behaviors) in caregivers based on SES (Medicaid vs. private insurance), and (4) explore correlations among context, process, and outcomes.

Methods

**Design, sample, setting and data collection procedures**

Participants for this cross-sectional descriptive study were caregivers of individuals with CF who were under the age of 18 and diagnosed with CF for at least 9 months. Caregivers were excluded if they were unable to read and write in English. This study was conducted at an academic
medical center with a pediatric CF center in the southeast region of the United States. Participants were approached during an outpatient clinic visit, consented and given a copy of the questionnaires. Participants had the opportunity to complete the questionnaires while at their visit or return them by mail.

Variables and measures
In the following section variables and measures will be organized by the conceptual framework.

Demographic and Clinical Characteristics of the Sample
Demographic information collected on the caregiver included age, gender, race, marital status, highest education completed, employment status, relationship to the child with CF, and information regarding the number of other children in the family. Demographic characteristics collected on the child with CF included gender, age and insurance status (used as a measure of SES). Clinical characteristics of the child included; lung function, height/weight/BMI percentile, and medications/treatments were obtained from the medical record.

Context Variables

Treatment complexity. Treatment complexity can include the number of drugs, route of administration, dosing frequency, and additional directions for taking the drug for example, relationship to timing with foods, set up of device to deliver drug (George, Phun, Bailey, Kong, & Stewart, 2004). The Treatment Complexity Scale (TCS) was developed by Sawicki et al. (2010) in an effort to evaluate CF treatment complexity. The TCS is a total score of 0-63, determined by assigning a complexity of 1, 2, or 3 for each therapy. The complexity score is based on daily frequency, duration, and ease of administration. The tool includes 37 chronic therapies that were captured as part of the longitudinal Epidemiology Study of Cystic Fibrosis (ESCF). Oral medications and metered-dose inhalers are assigned a score of 1 (20 items); once-
daily nebulized therapies and pancreatic enzyme are assigned a score of 2 (8 items), and airway clearances, twice-daily nebulized therapies, use of oxygen, continuous positive airway pressure (CPAP), bi-level positive airway pressure (BiPAP), and insulin or enteral supplements are assigned a score of 3 (9 items). A total score is calculated by summing the scores of all therapies. Higher scores indicate higher treatment complexity. Since the development of the tool, several new treatments for CF have been approved. The first is an oral medication that is taken twice a day. Similar to other twice a day medications this will be scored as 2. The second is a nebulized therapy, however, unlike other nebulized therapies which are administered once or twice daily, aztreonam is used 3 times a day. Although the treatment time is shorter (3-5 min compared to 10-20) it does require preparation and cleaning of nebulizer and will therefore receive a score of 3.

**Disease severity.** Disease severity was measured using information from pulmonary function testing. The pulmonary function testing was performed by the respiratory therapist as part of the routine clinic visit using the MCG diagnostics pulmonary function equipment with BreezeSuite© software (St. Paul MN). The forced expiratory volume in one second (FEV₁) was used and is classified into three groups: mild disease (FEV₁ >70%), moderate disease (FEV₁ 41–69%) and severe disease (FEV₁ <40%). This disease severity classification has been applied and adopted internationally in both the medical and psychosocial literature and is used as the standard for the epidemiological study in CF, and is the classification used in the Cystic Fibrosis Foundation (CFF) patient registry (Cystic Fibrosis Patient Registry, 2016 Annual Data Report). As an additional measure of disease severity, a visual analog scale (VAS) was used to gauge how the parent/caregiver viewed the child’s health. Participants were asked to rate their perception of the child’s health by placing an X on a 10cm line. One side of the line (0 cm) represented death
and the other end (10 cm) was perfect health. A score reflecting the position of the X on the line represented the perception of their child’s severity.

**Depression.** The Patient Health Questionnaire (PHQ-9) was used to screen for depression. In 2015, the International Committee on Mental Health in Cystic Fibrosis began recommending the use of this tool. The PHQ-9 is brief, reliable, and valid with appropriate cut off scores for detecting depressive symptoms. The score ranges from 0-27 with a recommended interpretation as follows: 1-4 minimal depression, 5-9 mild depression, 10-14 moderate depression, 15-19 moderately severe depression, and 20-27 severe depression. A cut off score of ≥ 10 has a sensitivity and specificity of 88% for major depression. Internal consistence is high with Chronbach’s alpha 0.86-.89 (Kroenke et al., 2001). In the present study, Cronbach’s alpha was 0.82.

**Process Variables**

**Self-efficacy.** To provide a broader assessment of self-efficacy, both general and disease-specific self-efficacy were considered. General self-efficacy was measured by the Perceived Health Competence Scale (PHCS). The PHCS contains 8 items to which responses are chosen from a 5 point Likert scale ranging from 1 = strongly disagree to 5 = strongly agree. There are 4 positively formulated items and 4 negatively formulated items. The four negatively worded items were re-coded so that a higher total score reflected self-efficacy (competence to manage their child’s health). The items add up to a total score with a range from 8-40; higher scores indicating higher levels of competence. The PHCS has been administered to wide variety of participants with a degree of health conditions. The psychometric properties suggest a high degree of internal consistency and satisfactory test-retest stability. Across five samples (young adult students, working adults, and persons with a chronic illness; rheumatoid arthritis) in the original study of
the PHCS there was high internal consistency with Cronbach’s alpha ranging from 0.82-0.90 (Smith, Wallson, Smith, 1995). The Cronbach’s alpha in this study was 0.70.

The Mountain West Cystic Fibrosis Consortium Questionnaire (MWCFC-Q) developed by McDonald, Christensen, Lingard, & Walker, (2009) was used to measure caregivers disease-specific self-efficacy. This Likert-type questionnaire (0=not at all confident to 10=completely confident) is divided into four sections 1) demographics, 2) food security, and 3) CF nutrition knowledge, and 4) confidence in CF management. In this study, only the 18 item section on confidence in CF management will be used. This section, which includes the three domains of CF care, allows the participants to rate their self-efficacy or confidence in each of these 3 domains: Task, Nutrition, CF Care center recommendations. Task measured the ability to manage CF care (pulmonary therapies and medications, nutrition addresses their ability to manage nutrition therapy and food related behaviors, and CF care center recommendations aimed to measure the participants perceived ability to follow the CF care center recommendations. The Cronbach alpha for the self-efficacy subscale has been 0.92 (McDonald et al, 2009), and in the present study it was 0.88.

Cystic fibrosis knowledge. CF knowledge was measured by the CF Knowledge and Attitudes Questionnaire developed by Siklosi, Gallagher and McKone (2010). The original questionnaire contains 49 fixed response questions, 2 open ended questions, and 9 questions combining both. The questionnaire is divided into three sections: demographics, CF knowledge test, and attitude and education questions. For the purpose of this study the CF knowledge test was used. The CF knowledge questionnaire is further divided into three sections: general knowledge (7 items), lung/GI issues (10 items), and reproductive/genetics knowledge (9 items). The CF knowledge test is scored by earning 1 point for each correct answer and 0 points for an
incorrect answer or the choice “I’m not sure”. A maximum score of 32 is possible. The
Cronbach’s alpha for the overall test has been reported as 0.75. The interclass correlation
coefficients ranged from 0.76-0.97 and indicate good test-retest reliability (Siklosi et al 2010).
The Cronbach’s alpha for the overall test was 0.80 in this sample.

Proximal Outcomes

**Family Self-Management of CF.** Self-management of CF was measured using The Self-
Management Questionnaire for Cystic Fibrosis (SMQ-CF) (Sockrider et al., 1996). This
instrument has both a caretaker and adolescent version, for this study the caretaker version was
used. The SMQ-CF measures behaviors that include skills characteristically used for
management of both pulmonary and gastrointestinal symptoms in CF. The SMQ-CF is a 46 item
written questionnaire using a 5 five point rating scale. Responses rank from 1 = never used to 5 =
always used. Items address behaviors reflecting monitoring and treatment of respiratory
infection, respiratory therapies, malabsorption and malnutrition. A ‘skip pattern’ is included for
patients who do not have pancreatic insufficiency and also for those who have not had an
experience with lower respiratory infections (bronchitis, bronchiolitis, or pneumonia). Due to
the “skip pattern”, each of the sections (respiratory surveillance, respiratory treatment, CPT/RT,
malabsorption, and nutritional surveillance) were examined separately based upon whether or not
the treatment was required for the child. For example, all caregivers responded to the respiratory
surveillance domain; however, if their child did not have an infection, the section on treatment
could be “skipped”. Limitations in determining validity exist as there is no “gold standard” for
comparison. Internal consistency was high with a Cronbach’s alpha coefficient of 0.95 for the
total scale (Sockrider et al, 1996). In the present study the overall Cronbach’s alpha was 0.87 and
the subscales were as follows: respiratory surveillance 0.94, respiratory treatment, 0.88, CPT/RT, 0.93, malabsorption, 0.81, and nutritional surveillance, 0.88.

Statistical analysis

All data were analyzed using SPSS (v. 24, Chicago, IL). Descriptive statistics were computed for sample demographics and all other variables using frequency distribution and percent for categorical variables and mean and SD based on distribution for continuous variables. Variables were inspected for normal distribution and means and SD were reported for those variables that were normally distributed. Medians values were provided for non-normally distributed values. Because the normal distribution assumption was not met, Mann-Whitney U test were conducted to compare insurance coverage groups (private, Medicaid) regarding depression, self-efficacy, knowledge, and family self-management behaviors. Pearson’s correlation coefficient ($r$) was used to determine relationships between context and process variables and self-management behaviors including treatment complexity, disease severity, knowledge, self-efficacy and depression.

Results

Sample

Thirty three individuals were invited to participate, 31 consented and 19 consented and returned questionnaires. There was not adequate data or either the parents or the child to make any comparisons between completers and non-completers. Sample characteristics for parents and caregivers are presented in Table 1. The average age of the caregiver participants was 40.71 years (SD=11.06 year; range = 25-69 years); 94% were women, 88.9% were Caucasian, and 80% were married. Most (87.5%) had at least one other biological child. The majority (81%) of
these participants had completed college or graduate school. Seventy-three percent of children had private insurance.

Characteristics of the child are included in Table 2. The majority of the children and adolescents were male (60%) with a mean age of 9.86 (SD=4.42 range= 1-16). The mean BMI percentile (%) was 49.83 (SD= 26.67 range= 3.46-96.75) suggesting that the BMI % for this cohort of children and adolescents is adequate. Additionally there was one participant under the age of 2; therefore, weight to length was utilized to characterize their nutritional status. The CF foundation recommends BMI % of 50% or greater. Five of these children with CF were also reported to have other health problems including 4 with asthma.

Descriptive statistics

Treatment complexity. Overall treatment complexity was 12.95 (SD=3.78, range 5-21). The mean treatment complexity in children ages 0-6 was 8, 7-13 years was 13.75 and 14-17 years was 14.25 suggesting that treatment complexity in this sample is substantial and increases with age. Standard therapies include nutritional support including vitamin and enzyme supplementation. Approximately 80% reported that their child received vitamin supplementation and 84% were on pancreatic enzyme replacement. Although the max score of this tool is significantly higher than the mean treatment complexity of this group it is important to put it into context to understand that a “lower” value could still be considered complex. For example a score of 10 could be representative of 12-15 pills, a twice a day inhaler, pancreatic enzymes 3-6 times a day, a nebulized medication treatment lasting 15 minutes/treatment (twice daily) and airway clearance lasting 20 minutes (twice daily).

Disease severity. The average FEV₁ percent predicted was 98.37 (SD= 15.83, range 66-120). Based on FEV₁ classification, the majority of these children (87.5%) had mild lung disease.
Caregivers’ perception of severity of their child’s disease severity (on the VAS) was 8.04 (SD=1.45, range= 4.5-9.9) indicating that their perception of the severity of disease was close to “perfect health”.

**Depression.** Overall, low rates of depressive symptoms were reported by the caregivers. The mean was 2.76 (SD=3.79 range 0-12), with a maximum score of 27 possible. Two participant’s scores represented mild depression and one participant score indicated moderate depression.

**Self-efficacy.** Both general and disease specific measures of the caregivers’ self-efficacy are reported in Table 3. The mean of general self-efficacy was 34.28 (SD=2.85; range 31-39) with a maximum possible score of 40. The CF-specific self-efficacy mean score was 163.89 (SD=16.81; range 122-180). The maximum score achievable is 180. Overall this represents moderate to high levels of confidence in ability to manage the child’s illness and treatment.

**Knowledge.** Knowledge scores, which were converted to a scale of 1-100 revealed an overall caregiver score of 70.64 (SD=15.32, range= 39.3-96.40) and subgroup scoring for general knowledge, lung/GI/ and reproductive/genetic were 65.7 (SD=21.1, range- 14.2-85.7), 81.48 (SD=12.65, range=58.3-100), 60.5 (SD=23.26, range= 11.1-100) respectively. Participants’ knowledge was greatest in the area of lung and GI information and the lowest in reproductive and genetic information.

**Self-management.** Self-management was divided into five subsections. Respiratory surveillance and treatment mean scores were 77.62 (SD=30.25; range= 0-100) and 87.19 (SD=11.47; range= 65.5-100) respectively. Self-management of chest and respiratory physical therapy had a mean of 72.65 (SD=31.27; range= 7.4-100). The subsections related to malabsorption and nutritional surveillance had a mean of 82.6 (SD=20.92; range 50-100) and
Participants’ self-management behaviors were greatest in respiratory treatment followed by malabsorption and nutritional surveillance. The lowest self-management behaviors were in the chest and respiratory therapy subsections.

Group Comparisons

Comparisons were made between participants with Medicaid vs. private insurance. Results are shown in Table 4. Overall participants with Medicaid had lower scores for total knowledge (including all sub categories), lower self-efficacy on both disease specific and generic scales, and higher depression ratings. A Mann Whitney U test was used to determine if the difference in the two medians were significantly different between groups. The results were significant for general knowledge (U=9.5, p=.038), reproductive and genetic knowledge (U=8, p=.019) and total knowledge (U=8, p=.026). There were no significant differences between groups with regards to depressive symptoms, lung specific knowledge, self-efficacy, or self-management behaviors.

Correlation analysis

Table 5 includes bivariate correlations between context and process variables and self-management behaviors represented by the model, including treatment complexity, disease severity, depressive symptoms, self-efficacy, and knowledge. There was one significant correlation identified between context factors (treatment complexity, perception of the child’s health status) and self-management. Caregivers’ perception of their child’s disease severity was negatively related to diet self-management behaviors (r= -.060, p=.008). This suggests that as the parent’s perception their child’s health improved, self-management behaviors related to nutritional surveillance were less of a priority. Although pancreatic sufficiency vs. insufficient information was not collected this may help explain this finding. Pancreatic sufficient patients
overall have improved health (perhaps reflected in caregivers’ perception), they also do not typically require the same level of diet management as pancreatic insufficient patients. There were significant correlations noted between process factors (self-efficacy, and knowledge) and self-management. General SE was negatively correlated with respiratory surveillance (r= -.490 p= 0.039). Conversely, disease specific SE was positively correlated with malabsorption (r= .637, p=.011) and nutritional surveillance components of self-management (r=.662 p= .003). Better knowledge of lung issues correlated with higher CPT/RT self-management (r=0.602 p=0.014) suggesting that in this case greater knowledge may positively affect behavior as it relates to CPT/RT. There were no significant correlations between treatment complexity or depressive symptoms and self-management.

Discussion

The majority of the caregiver participants were female and Caucasian. In one study of caregivers of children with CF, the mean age of caregivers was similar to the mean age of our sample (Driscoll, Montag-Leifling, Acton, Modi, 2009). The breakdown of male versus female caregivers our sample was significantly different; however, the authors (Driscoll et al) reported specifically seeking out male caregivers as they have been previously overlooked in research. The characteristics of the children in our study are similar to what is seen in the US population of CF patients. The CFF Registry data reports that the majority of individuals with CF are Caucasian (93.8%) and which was similar to our study population. We also had a similar gender breakdown with slightly more males than females. There were differences however seen between those with private insurance versus Medicaid. Our sample reported 73.7% private insurance whereas the Registry Data reported 59.5%, and Medicaid 44.8% versus our 26.3% (Registry Data 2015).
Treatment Complexity

The CFF maintains detailed clinical guidelines to aid the practitioner and individual/family with CF and includes various recommended treatments. Based on current CF chronic care guidelines, individuals with CF would be expected to be on multiple respiratory and non-respiratory treatments daily. In general, the sample appeared to be “following” the guidelines. Approximately 80% reported that their child received vitamin supplementation and 84% were on pancreatic enzyme replacement. Additionally, 100% of participants were prescribed chest physical therapy and/or respiratory therapy (CPT/RT). However, it is important to note that in order to fully assess if individuals are prescribed “appropriate” medications, a number of additional factors including: genotype, bacterial presence in the lungs, and CFRD status would have needed to be collected.

To date, there are limited published findings that used this measure of treatment complexity, the TCS. In the development of the TCS, Sawicki et al (2013) only included participants ages 6 and up. Ages were classified 6-13- child, 14-17 adolescent, and 18+ adults. In 2005, the mean TCS for child and adolescents were 12.4, and 12.6 respectively. The current study showed a mean of 8 in the under 6 year old group and in children between 6 and 13, a mean of 13.75, and in adolescents a mean of 14.25. These results were similar to the results published in 2013 reporting a mean TCS of 12.4 and 12.6 respectively. One possible explanation for the increase in scores could be the approval of two new Cystic Fibrosis transmembrane conductance regulators (CFTR), modifiers aimed at treating the underlying defect in CF. Ivacaftor/Kalydeco (potentiator of CFTR) and combination ivacaftor and lumacaftor/Orkambi (corrector of CFTR) are now considered standard of care for individuals with certain gene mutations. Forty-eight
percent of the children with CF in our sample were prescribed either ivacaftor or the combination of ivacaftor/lumacaftor resulting in an increase in treatment complexity of 2 points.

**Disease Severity**

The average FEV$_1$ in this group was high, with 6 of the 16 children having values exceeding 100% predicted. It is important to note that although FEV$_1$ is widely used in CF research it is not a sensitive measure of early lung disease. Our findings showed the majority of children with CF have mild impairment in pulmonary function.

Caregiver perception of child’s disease severity based on a VAS continuum from “death” to “perfect health” showed that overall perception trended towards “perfect health”. This positive perception suggests that caregivers are optimistic about their child’s health. One could speculate that the increasing life expectancy and the fairly recent approval of CFTR modulators may play a role. Additionally these children in this study are relatively “healthy” as demonstrated by high lung function, limited co-morbidities, and stable weights. To date there have been no other studies using the VAS in this manner however there are studies in other chronic diseases that health is positively perceived even in the face of chronic illness.

**Depression**

The findings of this study show that 16% of caregivers have symptoms of depression as evidence of scores greater than 5. We also found that the group with Medicaid had higher rates of depressive symptoms. Increased rates of depression in individual with chronic disease and their caregivers have been reported in the literature (Pinquart, & Shen, 2011). Results from a single center study report looking at depression in caregivers of CF found 20-35% increased rate of depression over the normal population (Latchford & Duff 2013). Driscoll et al. (2009) also found that 20-28% of female had increased depressive symptoms. In 2015, the CFF created new
care guidelines for the screening and treating depression and anxiety in people with CF and their caregivers. They recognized the need for a streamlined approach and recommended the use of the PHQ9 for screening (Quittner et al. 2015). By providing CF centers an easy to administer tool with specific guidelines on how to handle the results this streamline approach will hopefully lead to earlier identification of depressive symptoms, therefore earlier referrals and treatment. It will also be helpful as individuals with CF transfer to other pediatric CF centers or transition to adult centers as there will be a record of baseline depressive symptoms for comparison.

**Self-efficacy**

The findings of this study showed that both general and disease specific self-efficacy were relatively high in this population. Although the overall disease specific self-efficacy was high there were several areas identified that participants were less confident. These related to how confident they were in managing feeding/eating/mealtime behavior issues in their child with CF. The other area that was identified was the ability to manage their child’s CF in such a way that emotional distress does not affect everyday life. While self-efficacy has been explored in detail in several chronic conditions, it has not yet been fully explored in CF. However, chronic disease literature has demonstrated that it is crucial for caregivers of children with CF are confident in their ability to manage the complexities of the treatment. Therefore, studies to examine the role of caregiver self-efficacy should be considered.

**Knowledge**

Findings of this study were consistent with previous research that identified possible knowledge gaps-- specifically related to reproduction and genetics. Our findings indicated, overall, that participants’ knowledge of lung and GI topics (physiotherapy, lung infections, and malabsorption) was greatest, and the lowest in reproductive health and genetics topics (fertility
and recurrence risk). Siklosi et al. (2010) found that although adult CF patients understood most aspects of their disease, knowledge deficits related to genetics and reproduction were identified. These same researchers also reported significant associations between knowledge scores and gender and education attainment but did not find significant differences with SES. Being female and having higher education attainment were both independent predictors of higher CF knowledge scores. In the current study, group comparisons were made between individuals with private or Medicaid. Our findings showed that participants with Medicaid had lower knowledge scores. Gage (2012) also found that both males and females lack disease specific reproductive health knowledge and further identified the need for specific sexual and reproductive education. As the CF population continues to live longer and healthier lives it will be critical for improvements in knowledge regarding genetics and reproduction.

Self-management

The management of CF is more complex than just implementing prescribed therapies; it requires patients and families to monitor symptoms and make decisions to modify therapies appropriately. Therefore, skills related to monitoring, decision-making, communicating, and coping are necessary to enable patients and families to perform self-management behaviors (Parcel et al., 1994). Previous research has demonstrated that individuals with CF are able to perform 30-70% of these self-management tasks (Abbot, Dodd, & Webb, 1996). Patients with CF, however, have more difficulty performing some treatments than others with the highest percent of self-management behaviors in antibiotics management and the lowest in physiotherapy behaviors (Abbot et al., 1996). The results of this study found that self-management was greatest with regards to respiratory and nutritional surveillance. This is not surprising for several reasons. First both respiratory and nutrition are the major focus areas of
care in this population. Secondly both are able to be measured (lung function, BMI) and are used as “benchmarks” for how well an individual with CF is doing. Finally, the leading cause of death in this population is directly related to respiratory health and nutrition.

Limitations

There are several limitations in the study, most notably the small sample size. There were 12 caregivers who consented but never returned the questionnaires which may have led to potential selection bias. Interestingly of the questionnaires that were not returned the majority did not have FEV1 percent predicted results which indicate a younger age of the child with CF, this could possibly suggest limited time for parents of younger children to complete surveys. Often it was the caregiver of younger children who were unable to complete the questionnaire in the clinic because they were busy taking care of the younger child during the clinic visit. In addition, the study was limited by self report data which can lead to overestimation of factors (e.g., self-efficacy, self-management) and underestimation of depressive symptoms. Furthermore, in developing this study it was recognized that the disease specific measure of self-management in the CF population was over 20 years old and had limited use. Therefore this measure may not have fully captured updated practice recommendations and treatment guidelines. Generalizability was limited because of the single site data collection and majority participation from female caregivers.

Summary & Conclusions

This was the first time that the IFMST was used to guide CF research. The IFMST would provide a consistent framework to guide future studies aimed at identifying factors that influence self-management behaviors of CF in patients and their caregivers. Overall this study of primarily female caregivers was described as having high SE, and average knowledge. Deficits were noted
in the areas of reproductive and genetic knowledge. This group overall felt like their children with CF was close to perfect health. The children with CF in this study had moderate treatment complexity and high lung function. This study also found differences between Medicaid and private insurance groups related to knowledge. There were also significant relationships between disease severity and CF specific SE and nutritional surveillance as well as general SE and respiratory surveillance.

**How Might This Information Affect Nursing Practice**

The results of this study have several implications for nursing practice. Because of the knowledge gaps identified, clinicians need to continually reinforce caregivers understanding of the disease and in particular with regards to reproductive and genetics knowledge, although it is important to note that knowledge alone is not sufficient to improve self-management. Self-management plays an important role in successful care of chronic diseases including CF and it is critical to expand the understanding of what possible factors help predict or drive self-management so that appropriate interventions can be implemented. It is also critical to continue to create tools that are relevant, reliable and valid to assist in the measurement of self-management. Additionally results from this study can help guide future research in CF as well as other rare diseases. For the first time ever the number of adult’s with CF is greater than the number of children. Although this is a testament to the hard work of scientist, researchers, clinicians, and patients it can pose potential limitations on the number of available participants in the pediatric clinics. Smaller numbers coupled with competing research studies emphasizes the need for multiple sites. Additionally, due to space and time constraints in the clinic setting it would be useful to employ an electronic method of data collection.
Table 1: Sample Characteristics of Parent/Caregiver

<table>
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<th>Variables</th>
<th>Frequency</th>
<th>Mean (SD)</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Contextual Factors</strong></td>
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</tr>
<tr>
<td>Demographics</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Age, y (n=17)</td>
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<td>40.71 (11.06)</td>
<td>25-69</td>
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<tr>
<td>25-35</td>
<td>5</td>
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</tr>
<tr>
<td>36-45</td>
<td>6</td>
<td>(37.5)</td>
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</tr>
<tr>
<td>46-55</td>
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<td>(25)</td>
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</tr>
<tr>
<td>Older than 56</td>
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<td>(6.3)</td>
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<tr>
<td>Gender (n=18)</td>
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<td></td>
<td></td>
</tr>
<tr>
<td>Women</td>
<td>17</td>
<td>(94.4)</td>
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</table>
### Physical & Social Environment

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<th>Ethnicity  (n=18)</th>
<th>Frequency</th>
<th>%</th>
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<tr>
<td>Non-Hispanic</td>
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<tr>
<td>Hispanic</td>
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<tr>
<td>Race (n=18)</td>
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<tr>
<td>White</td>
<td>16 (88.9)</td>
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<tr>
<td>Black</td>
<td>2 (11.1)</td>
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### Individual & Family Factors

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<th>Marital Status (n=15)</th>
<th>Frequency</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Married/Life Partner</td>
<td>12 (80)</td>
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</tr>
<tr>
<td>Divorced/Separated</td>
<td>2 (13.3)</td>
<td></td>
</tr>
<tr>
<td>Single/Never Married</td>
<td>1 (6.7)</td>
<td></td>
</tr>
<tr>
<td>Relationship to Child with CF (n=16)</td>
<td>Frequency</td>
<td>%</td>
</tr>
<tr>
<td>Mother</td>
<td>14 (87.5)</td>
<td></td>
</tr>
<tr>
<td>Maternal Grandmother</td>
<td>1 (6.3)</td>
<td></td>
</tr>
<tr>
<td>Father</td>
<td>1 (6.3)</td>
<td></td>
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<tr>
<td>Education (n=16)</td>
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<td></td>
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<tr>
<td>≤ High School</td>
<td>1 (6.3)</td>
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</tr>
<tr>
<td>Completed High School</td>
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</tr>
<tr>
<td>Started College/Tech School</td>
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<tr>
<td>Completed College</td>
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<tr>
<td>Completed Graduate School</td>
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<td>Part time</td>
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<td>Out of work &gt; 1 year</td>
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<td>Homemaker</td>
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<td>Retired</td>
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<tr>
<td>Other Children (n=16)</td>
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<td></td>
</tr>
<tr>
<td>Yes</td>
<td>14 (87.5)</td>
<td></td>
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<tr>
<td>No</td>
<td>2 (12.5)</td>
<td></td>
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<tr>
<td>Number of Other Children (n= 13)</td>
<td>Frequency</td>
<td>%</td>
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<tr>
<td>1</td>
<td>6 (46.2)</td>
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<td>2</td>
<td>5 (16.7)</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>2 (15.4)</td>
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**Table 2: Child Characteristics**
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<thead>
<tr>
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<th>Range</th>
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<tbody>
<tr>
<td>Height, cm (n=19)</td>
<td>131.09 (23.64)</td>
<td>77-163.6</td>
</tr>
<tr>
<td>Weight, kg</td>
<td>33.17 (13.76)</td>
<td>11.05-58.50</td>
</tr>
<tr>
<td>BMI %</td>
<td>49.83 (26.67)</td>
<td>3.46-96.75</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Males</td>
<td>15 (60%)</td>
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</tr>
<tr>
<td>Treatment Complexity</td>
<td>12.95 (3.78)</td>
<td>5-12</td>
</tr>
<tr>
<td>FEV₁ (n=16)</td>
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<td></td>
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<tr>
<td>FEV₁ % Predicted</td>
<td>98.37 (15.83)</td>
<td>66-120</td>
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<tr>
<td>FEV₁ Classification</td>
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<td></td>
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<tr>
<td>• Mild</td>
<td>14 (87.5)</td>
<td></td>
</tr>
<tr>
<td>• Moderate</td>
<td>2 (12.5)</td>
<td></td>
</tr>
<tr>
<td>• Severe</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>Age</td>
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<td>1-16</td>
</tr>
<tr>
<td>Years Diagnosed</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• 0-5</td>
<td>4 (23.5)</td>
<td></td>
</tr>
<tr>
<td>• 6-10</td>
<td>4 (23.5)</td>
<td></td>
</tr>
<tr>
<td>• 11-15</td>
<td>8 (47)</td>
<td></td>
</tr>
<tr>
<td>• &lt; 16</td>
<td>1 (6)</td>
<td></td>
</tr>
<tr>
<td>Other Health Problems</td>
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<tr>
<td>• Yes</td>
<td>5 (29.4)</td>
<td></td>
</tr>
<tr>
<td>• No</td>
<td>12 (70.6)</td>
<td></td>
</tr>
<tr>
<td>Insurance (n=19)</td>
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<td></td>
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<tr>
<td>Private Insurance</td>
<td>14 (73.7)</td>
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</tr>
<tr>
<td>Medicaid</td>
<td>5 (26.3)</td>
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</tbody>
</table>

Table 3: *Descriptive Statistics of Model Variables for Parent/Caregiver*
### Context

<table>
<thead>
<tr>
<th>Condition-Specific</th>
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</thead>
<tbody>
<tr>
<td>Disease Severity</td>
</tr>
<tr>
<td>• Perception</td>
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*Individual and Family*

| Depressive Symptoms | N=18 | 2.76 (3.78) | 0-12 |

*Process*

*Knowledge and Beliefs*

<table>
<thead>
<tr>
<th>Knowledge</th>
<th>N=17</th>
<th>70.64 (15.32)</th>
<th>39.3-96.4</th>
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</thead>
<tbody>
<tr>
<td>• Total</td>
<td></td>
<td>65.70 (21.1)</td>
<td>14.3-87.5</td>
</tr>
<tr>
<td>• General</td>
<td></td>
<td>81.48 (12.65)</td>
<td>58.3-100</td>
</tr>
<tr>
<td>• Lung</td>
<td></td>
<td>60.5 (23.26)</td>
<td>11.1-100</td>
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<td>• Reproductive</td>
<td></td>
<td></td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Self Efficacy</th>
<th>N=18</th>
<th>34.28 (2.85)</th>
<th>31-39</th>
</tr>
</thead>
<tbody>
<tr>
<td>• General</td>
<td></td>
<td>163.89 (16.81)</td>
<td>122-180</td>
</tr>
<tr>
<td>• Disease Specific</td>
<td></td>
<td></td>
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</table>

*Outcome*

*Family Self Management Behaviors*

<table>
<thead>
<tr>
<th>Self Management*</th>
<th>N=18</th>
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<th>0-100</th>
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</thead>
<tbody>
<tr>
<td>• Respiratory Surveillance</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Respiratory Treatment</td>
<td>N=10</td>
<td>87.19 (11.47)</td>
<td>65.5-100</td>
</tr>
<tr>
<td>• Chest Physical and Respiratory therapy</td>
<td>N=17</td>
<td>72.65 (31.27)</td>
<td>7.4-100</td>
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<tr>
<td>• Malabsorption</td>
<td>N=14</td>
<td>82.16 (20.92)</td>
<td>50-100</td>
</tr>
<tr>
<td>• Nutritional Surveillance</td>
<td>N=17</td>
<td>82.55 (17.78)</td>
<td>34.4-100</td>
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</table>

<table>
<thead>
<tr>
<th>Insurance</th>
<th>Median</th>
<th>U</th>
<th>P</th>
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*Tool allowed for a “skip pattern” for sections that were N/A*

---

**Table 4: Group Comparisons between Medicaid and Private Insurance**

61
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<tr>
<th>Category</th>
<th>Medicaid</th>
<th>Private</th>
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<th>p</th>
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<td>71.4</td>
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<td>.038</td>
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<td>Medicaid</td>
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<td>78.6</td>
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<td>168</td>
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<td>Perceived Health Competence Scale</td>
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<td>58.3</td>
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<td>91.5</td>
<td>22.5</td>
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<td>Malabsorption Self Management Behaviors</td>
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<td>75.1</td>
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<td>23.5</td>
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Table 5: Correlations between Context and Process Variables and Self Management Behaviors

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<th>Context:</th>
<th>Outcomes:</th>
<th>Respiratory Surveillance</th>
<th>Respiratory Treatment</th>
<th>CPT/RT</th>
<th>Malabsorption</th>
<th>Nutritional Surveillance</th>
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<tbody>
<tr>
<td>• Treatment Complexity</td>
<td>Pearson Correlation</td>
<td>.082</td>
<td>.476</td>
<td>.275</td>
<td>-.185</td>
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<td></td>
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<td>.746</td>
<td>.174</td>
<td>.285</td>
<td>.590</td>
<td>.665</td>
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<tr>
<td></td>
<td>N</td>
<td>18</td>
<td>9</td>
<td>17</td>
<td>14</td>
<td>17</td>
</tr>
<tr>
<td>• Perceived Disease Severity</td>
<td>Pearson Correlation</td>
<td>.377</td>
<td>-.421</td>
<td>.008</td>
<td>.042</td>
<td>-.606**</td>
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<td>.123</td>
<td>.209</td>
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<td>.872</td>
<td>.008</td>
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<td>18</td>
<td>9</td>
<td>17</td>
<td>15</td>
<td>17</td>
</tr>
<tr>
<td>• Depression</td>
<td>Pearson Correlation</td>
<td>.373</td>
<td>.129</td>
<td>-.008</td>
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<td>.127</td>
<td>.732</td>
<td>.975</td>
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<td></td>
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<td>10</td>
<td>17</td>
<td>15</td>
<td>17</td>
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<td>Process:</td>
<td>General Knowledge</td>
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<td>-.243</td>
<td>-.309</td>
<td>-.004</td>
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<td>.437</td>
<td>.419</td>
<td>.988</td>
<td>.402</td>
<td>.525</td>
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<td>17</td>
<td>9</td>
<td>16</td>
<td>13</td>
<td>16</td>
</tr>
<tr>
<td>• Lung/GI Knowledge</td>
<td>Pearson Correlation</td>
<td>.316</td>
<td>-.054</td>
<td>.602*</td>
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<td>.286</td>
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<tr>
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<td>.889</td>
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<td>.875</td>
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<td>16</td>
<td>13</td>
<td>16</td>
</tr>
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<td>• Genetic/Reproductive Knowledge</td>
<td>Pearson Correlation</td>
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<td>-.451</td>
<td>.285</td>
<td>.460</td>
<td>.220</td>
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<td>.223</td>
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<td>.881</td>
<td>.936</td>
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<td>9</td>
<td>16</td>
<td>13</td>
<td>16</td>
</tr>
<tr>
<td>• Total Knowledge</td>
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<td>.51</td>
<td>-.323</td>
<td>.337</td>
<td>-.067</td>
<td>.055</td>
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<tr>
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<td>Significance</td>
<td>.845</td>
<td>.397</td>
<td>.202</td>
<td>.828</td>
<td>.839</td>
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<td></td>
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<td>9</td>
<td>16</td>
<td>13</td>
<td>16</td>
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<tr>
<td>• General SE</td>
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<td>-.592</td>
<td>-.093</td>
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<td>.039</td>
<td>.071</td>
<td>.723</td>
<td>.200</td>
<td>.788</td>
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<td>18</td>
<td>10</td>
<td>17</td>
<td>14</td>
<td>17</td>
</tr>
<tr>
<td>• CF Specific SE</td>
<td>Pearson Correlation</td>
<td>.190</td>
<td>.117</td>
<td>.311</td>
<td>.637*</td>
<td>.662**</td>
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<td>.011</td>
<td>.003</td>
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<td>19</td>
<td>10</td>
<td>18</td>
<td>15</td>
<td>18</td>
</tr>
</tbody>
</table>

Note: N ≥18 reflected a skip pattern on the self management behavior tool
References


“About CF.” *About CF*, Cystic Fibrosis Foundation, retrieved from [http://www.cff.org/AboutCF](http://www.cff.org/AboutCF)

Cystic Fibrosis Foundation Patient Registry, 2016 Annual Data Report. Bethesda, Maryland

Cystic Fibrosis Foundation.


Chapter 5
DISCUSSION OF FINDINGS

The purpose of this study was to explore context, process, and outcomes using the IFSMT as a guide. The findings were presented in detail in Chapter 4. This chapter will address the limitations of the research, implications for practice, theory, and future research.

Limitations

There are several limitations in the study, notably the small sample size. In additional the study was limited by self report data which can lead to overestimation of factors (self efficacy, self management) and underestimation of depression. Generalizability was also limited because of the single site data collection and majority participation from females.

Recruitment was affected by a number of factors including competing research projects in the clinic, nature of multi-disciplinary clinics, space limitations and lack of additional sites for enrollment. Over the one year data collection period there were several other research opportunities occurring at the same time. One study in particular required a pretest, a 30 minute educational session and a posttest the following clinic visit. This greatly impacted the ability for possible participants to participate in an additional study for a 6 month time frame. Additionally the clinic participates in multiple other research studies, including clinical trials, as well as quality improvement projects that limited the ability to participate. Attempts were made to provide additional options for participation including an option to return the surveys by mail. Participants were given a prepaid self addressed envelope to return the surveys. Unfortunately, although this worked for some participants, there were an additional 12 participants, including 3 male participants, which consented but failed to return the surveys.
One other area that posed a problem was the number of providers who needed to see each patient during a clinic visit. The clinic where data collection took place was a multi-disciplinary clinic. The clinical team included pulmonary physicians (including fellows, residents, and medical students), endocrine physicians, respiratory therapy, nutrition, social work, pharmacy, psychology, research staff, and nursing. During the pre clinic meeting each individual with CF was discussed and it was noted which team members needed to which person. It was not uncommon for up to 6-10 people to need to meet with the individual and their family.

Space in the clinic was an additional challenge. Due to infection control requirements, there are specific rooms in the clinic designated to the CF clinic. The CF clinic has 5 negative pressure rooms that allow for back to back CF individuals without a “washout” time. When these rooms were full then other space in the clinic could be used but required a specified time frame between patients. Essentially in the 1-2 hour appointment all of the various specialties were trying to get in to see the individual and family. There was a consult room that was available, however after a several hour appointment the patients and families were ready to leave clinic to return to their daily activities.

The original proposal included a second site for data collection. Bon Secours Hospital (St. Mary’s Campus) had a small CF clinic which treated 10-20 eligible individuals. However, due to the unexpected retirement, and subsequent departure of an additional pulmonary physician, the site was no longer available for data collection. This was unfortunate because unlike the primary site this site had limited to no research and fewer barriers with regards to time and space.
Implications for Nursing Practice

The results of this study have several implications for nursing practice. Because of the knowledge gaps identified, clinicians need to continually reinforce caregivers understanding of the disease and in particular with regards to reproductive and genetics knowledge, although it is important to note that knowledge alone is not sufficient to improve self-management. Self-management plays an important role in successful care of chronic diseases including CF and it is critical to expand the understanding of what possible factors help predict or drive self-management so that appropriate interventions can be implemented. It is also critical to continue to create tools that are relevant, reliable and valid to assist in the measurement of self-management. Additionally, results from this study can help guide future research in CF as well as other rare diseases. For the first time ever, the number of adult’s with CF is greater than the number of children. Although this is a testament to the hard work of scientist, researchers, clinicians, and patients, it can pose potential limitations on the number of available participants in the pediatric clinics. Smaller numbers coupled with competing research studies emphasize the need for multiple sites. Additionally, due to space and time constraints in the clinic setting, it would be useful to employ an electronic method of data collection.

Implications for Nursing Theory and Research

This study was the first time the IFMST was used to examine the CF population. The results provided evidence that the theory provides an organized method to examining factors that relate to self-management behaviors. More research with larger samples and multiple sites are needed to further explore the role of the IFMST in the CF population. Future studies using the IFMST in families should consider family structure and functioning to better understand this factor.
Summary

Over the past 20 years little research has been conducted on factors related to self-management in the CF populations. Additional those studies that have explored factors have been intervention focused without first adequately describing the baseline factor. This was the first time that the IFMST was used to guide CF research. The IFMST would provide a consistent framework to guide future studies aimed at identifying factors that influence self-management behaviors of CF in patients and their caregivers. Overall this study of primarily female caregivers was described as having high SE, and average knowledge. Deficits were noted in the areas of reproductive and genetic knowledge. This group overall felt like their children with CF was close to perfect health. The children with CF in this study had moderate treatment complexity and high lung function. This study also found differences between Medicaid and private insurance groups related to knowledge. There were also significant relationships between disease severity and CF specific SE and nutritional surveillance as well as general SE and respiratory surveillance.
REFERENCES
REFERENCES


“About CF.” About CF, Cystic Fibrosis Foundation, retrieved from http://www.cff.org/AboutCF

Cystic Fibrosis Foundation Patient Registry, 2016 Annual Data Report. Bethesda, Maryland

Cystic Fibrosis Foundation.


APPENDIX A

Demographic Questionnaire
DEMOGRAPHIC QUESTIONNAIRE

These questions are about YOU—not your child. Please read carefully and mark your response(s).

1. What is your current age? ______

2. What is your gender?
   □ Male
   □ Female

3. What is your ethnicity?
   □ Hispanic or Latino
   □ Not Hispanic or Latino

4. What is your race?
   □ American Indian or Alaska Native   □ Hawaiian or Other Pacific Islander
   □ Asian   □ Black or African American
   □ White

5. What is your highest educational level?
   □ Some grammar school completed. What grade? ________
   □ Did not complete high school
   □ Completed high school
   □ Started college or technical school
   □ Completed technical school
6. What is your employment status? Check all that apply.

☐ Employed full-time for wages
☐ Employed part-time for wages
☐ Out of work for more than 1 year
☐ Out of work for less than 1 year
☐ A homemaker ☐ A student
☐ Retired ☐ Unable to work; receiving disability

7. What is your marital status?

☐ Married/ Life partner ☐ Divorced/Separated ☐ Widowed
☐ Single, never been married ☐ Other: ____________________

8. What is your relationship to the child with CF?

☐ Mother ☐ Maternal Grandmother ☐ Maternal Grandfather
☐ Father ☐ Paternal Grandmother ☐ Paternal Grandfather
☐ Other (please list) : ____________________

9. In addition to your child with CF do you have other biological children?

☐ Yes
☐ No

If yes, please list ages: __________________________________________________
10. In addition to your child with CF do you have other non-biological children?

☐ Yes

☐ No

If yes, please list ages: ____________________________________________________________

The following questions are about your child with CF

11. What is the age of your child with Cystic Fibrosis? ______

12. How many years has your child been diagnosed with Cystic Fibrosis? ______

13. Has your child ever been treated in another CF center?

☐ Yes

☐ No

14. Does your child have any other health problems (either related to or not related to CF)?

☐ Yes

☐ No

If yes, please list:
___________________________________________________________
___________________________________________________________
___________________________________________________________
APPENDIX B

Treatment Complexity Score
## TREATMENT COMPLEXITY SCORE

<table>
<thead>
<tr>
<th>TCS Score = 1 point</th>
<th>TCS Score = 2 points</th>
<th>TCS Score = 3 points</th>
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</thead>
<tbody>
<tr>
<td>Inhaled bronchodilator</td>
<td>Hypertonic saline</td>
<td>Airway clearance technique</td>
</tr>
<tr>
<td>Oral bronchodilator</td>
<td>N-acetyl cysteine (Mucomyst)</td>
<td>CPAP/BIPAP</td>
</tr>
<tr>
<td>Inhaled corticosteroid</td>
<td>Pancreatic enzymes</td>
<td>Oxygen</td>
</tr>
<tr>
<td>Oral corticosteroid</td>
<td>Inhaled TOBI (once per day)</td>
<td>Enteral supplement</td>
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<tr>
<td>High dose ibuprofen</td>
<td>Inhaled other aminoglycoside (once per day)</td>
<td>Parenteral supplement</td>
</tr>
<tr>
<td>Leukotriene inhibitor/antagonist</td>
<td>Inhaled colistin (once per day)</td>
<td>Inhaled TOBI (twice per day)</td>
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<td>Cromolyn/mast cell stabilizer</td>
<td>Other inhaled chronic suppressive antibiotic</td>
<td>Inhaled other aminoglycoside (twice per day)</td>
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<td>Gastric acid suppressors</td>
<td>Dornase alfa</td>
<td>Inhaled colistin (twice daily)</td>
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<td>CFTR modulators</td>
<td>Insulin</td>
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<td>Ursodiol</td>
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<td>Cayston</td>
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<tr>
<td>Vitamins</td>
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<td>Oral quinolone</td>
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<td>Contraceptive/hormonal methods</td>
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<td>Diuretic</td>
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</table>
APPENDIX C

Visual Analog Scale
VISUAL ANALOG SCALE

Please draw a line (I) on how you feel about your child’s current health.

Death

Perfect Health
APPENDIX D

The Patient Health Questionnaire (Phq-9)
THE PATIENT HEALTH QUESTIONNAIRE (PHQ-9)

Over the past 2 weeks, how often have you been bothered by any of the following problems?  

<table>
<thead>
<tr>
<th></th>
<th>Not at All</th>
<th>Several Days</th>
<th>More Than Half the Days</th>
<th>Nearly Every Day</th>
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</thead>
<tbody>
<tr>
<td>1. Little interest or pleasure in doing things</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>2. Feeling down, depressed or hopeless</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>3. Trouble falling asleep, staying asleep, or sleeping too much</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>4. Feeling tired or having little energy</td>
<td>0</td>
<td>1</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>5. Poor appetite or overeating</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>6. Feeling bad about yourself- or that you’re a failure or have let yourself or your family down</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>7. Trouble concentrating on things, such as reading the newspaper or watching television</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>8. Moving or speaking so slowly that other people could have noticed. Or the opposite-being so fidgety or restless that you have been moving around a lot more than usual</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>9. Thoughts that you would be better off dead or of hurting yourself in some way</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
</tr>
</tbody>
</table>
APPENDIX E

Perceived Health Competence Scale
**PERCEIVED HEALTH COMPETENCE SCALE**

1. I handle myself well with respect to my (child’s) health

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>

2. No matter how hard I try, my (child’s) health just doesn’t turn out the way I would like.

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>

3. It is difficult for me to find effective solutions to the health problems that come my (child’s) way.

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>

4. I succeed in the projects I undertake to improve my (child’s) health.

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>

5. I am generally able to accomplish my goals with respect to (my child’s) health.

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>

6. I find my efforts to change things I don’t like about my (child’s) health are ineffective.

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>

7. Typically, my plans for my (child’s) health don’t work out well.

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>

8. I am able to do things for my (child’s) health as well as most other people.

<table>
<thead>
<tr>
<th></th>
<th>1 Strongly Disagree</th>
<th>2 Disagree</th>
<th>3 Neutral</th>
<th>4 Agree</th>
<th>5 Strongly Agree</th>
</tr>
</thead>
</table>
APPENDIX F

Mountian West Cystic Fibrosis Consortium Questionnaire
Mountain West Cystic Fibrosis Consortium Questionnaire

Important: the daily care for a child with CF involves many different tasks. Parents must oversee, or manage, these tasks to make sure they are done as prescribed. We would like to know how confident you feel in managing certain areas of your child’s CF care.

Managing your child’s CF treatments, foods, or mediation does NOT mean that you personally do every treatment or give every medication. A manager may assign extended family, daycare, babysitters, school or even the child with CF some of the task necessary for CF care.

For each of the following questions, please circle the number that describes your confidence that you can managed the task described on a regular basis at the present time.

1. How confident are you that you can correctly perform all the tasks and activities to manage your child’s CF care?

   1 Not at all confident
   2 Slightly confident
   3 Quite confident
   4 Completely confident

2. How confident are you that you can tell when your child with CF needs to be seen for a sick visit to the CF clinic or doctor’s office before a problem becomes an emergency?

   1 Not at all confident
   2 Slightly confident
   3 Quite confident
   4 Completely confident
3. How confident are you that you can manage your child’s CF at home in such a way that your child does not need to go as often to the CF clinic, doctor’s office, or emergency room for sick visits?

1 2 3 4 5 6 7 8 9 10
Not at all confident Slightly confident Quite confident Completely confident

4. How confident are you that you can correctly manage the respiratory treatments that have been prescribed for your child with CF?

1 2 3 4 5 6 7 8 9 10
Not at all confident Slightly confident Quite confident Completely confident

5. How confident are you that you can manage any inhaled medications (such as Pulmozyme or Tobi) that have been prescribed for your child with CF?

1 2 3 4 5 6 7 8 9 10
Not at all confident Slightly confident Quite confident Completely confident

6. How confident are you that you can manage any oral medications (such as enzymes or antibiotics) for your child with CF?

1 2 3 4 5 6 7 8 9 10
Not at all confident Slightly confident Quite confident Completely confident
7. How confident are you that you can manage the vitamin supplements prescribed for your child?

1  2  3  4  5  6  7  8  9  10
Not at all confident  Slightly confident  Quite confident  Completely confident

8. How confident are you that you could manage tube feedings for your child with CF if they were prescribed?

1  2  3  4  5  6  7  8  9  10
Not at all confident  Slightly confident  Quite confident  Completely confident

9. Having a child with a chronic illness can cause emotional distress within a family. How confident are you that you can manage your child’s CF in such a way that emotional distress does not affect your everyday life?

1  2  3  4  5  6  7  8  9  10
Not at all confident  Slightly confident  Quite confident  Completely confident

10. How confident are you that you can manage the weight gain for your child with CF to achieve the best growth possible?

1  2  3  4  5  6  7  8  9  10
Not at all confident  Slightly confident  Quite confident  Completely confident
11. How confident are you that you understand the weight gain and calorie intake goals recommended by your child’s CF center?

<table>
<thead>
<tr>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
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<th>6</th>
<th>7</th>
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<th>10</th>
</tr>
</thead>
<tbody>
<tr>
<td>Not at all confident</td>
<td></td>
<td></td>
<td>Slightly confident</td>
<td></td>
<td>Quite confident</td>
<td></td>
<td></td>
<td></td>
<td>Completely confident</td>
</tr>
</tbody>
</table>

12. How confident are you that you can manage the special diet required for your child with CF?

<table>
<thead>
<tr>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
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<th>9</th>
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</tr>
</thead>
<tbody>
<tr>
<td>Not at all confident</td>
<td></td>
<td></td>
<td>Slightly confident</td>
<td></td>
<td>Quite confident</td>
<td></td>
<td></td>
<td></td>
<td>Completely confident</td>
</tr>
</tbody>
</table>

13. How confident are you that you can manage the diet or nutritional needs for other family members as well as for those for the child with CF?

<table>
<thead>
<tr>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
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</tr>
</thead>
<tbody>
<tr>
<td>Not at all confident</td>
<td></td>
<td></td>
<td>Slightly confident</td>
<td></td>
<td>Quite confident</td>
<td></td>
<td></td>
<td></td>
<td>Completely confident</td>
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</tbody>
</table>

14. How confident are you that you are able to manage any feeding, eating or mealtime behavior issues for your child with CF?

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<tbody>
<tr>
<td>Not at all confident</td>
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<td></td>
<td>Slightly confident</td>
<td></td>
<td>Quite confident</td>
<td></td>
<td></td>
<td></td>
<td>Completely confident</td>
</tr>
</tbody>
</table>
15. How confident are you that taking your child to CF center visits at least every three months helps your child, even when your child is doing well?

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<thead>
<tr>
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<th>1</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Not at all confident</td>
<td>Slightly confident</td>
<td>Quite confident</td>
<td>Completely confident</td>
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</table>

16. How confident are you that your CF center has provided you with best information available to help your child with CF?

<table>
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</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Not at all confident</td>
<td>Slightly confident</td>
<td>Quite confident</td>
<td>Completely confident</td>
<td></td>
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</tbody>
</table>

17. Overall, how confident are you that if you do all the treatments, medication and recommendations from the CF center, your child with CF will live a longer, healthier life?

<table>
<thead>
<tr>
<th></th>
<th>1</th>
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<th>4</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Not at all confident</td>
<td>Slightly confident</td>
<td>Quite confident</td>
<td>Completely confident</td>
<td></td>
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</tbody>
</table>

18. Overall, how confident are you that you are able to follow all the recommendations from the CF center?

<table>
<thead>
<tr>
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<th>4</th>
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<th>7</th>
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<th>9</th>
<th>10</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Not at all confident</td>
<td>Slightly confident</td>
<td>Quite confident</td>
<td>Completely confident</td>
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</tbody>
</table>
APPENDIX G

CF Knowledge and Attitudes Questionnaire
CF KNOWLEDGE AND ATTITUDES QUESTIONNAIRE

CF Knowledge- General Knowledge and complications

1. What is the basic problem in the body that causes symptoms of CF?
   - □ The body has severe allergies to food and particles in the air
   - □ The body has a weak immune system
   - □ The cells in the body cannot properly regulate salt
   - □ Several organs in the body are underdeveloped
   - □ I’m not sure

2. Do people with CF sweat more, less, or the same amount as people without CF?
   - □ More
   - □ Less
   - □ The same amount
   - □ I’m not sure

3. Do people with CF have more, less, or the same amount of salt in their sweat as people without CF?
   - □ More
   - □ Less
   - □ The same amount
   - □ I’m not sure

4. Do people with CF have higher, lower, or the same chance of developing diabetes as people without CF?
   - □ Higher
   - □ Lower
   - □ The same
   - □ I’m not sure

5. Do people with CF have higher, lower, or the same chance of developing liver disease as people without CF?
   - □ Higher
   - □ Lower
   - □ The same
   - □ I’m not sure

6. Some people with CF develop kidney disease. What is the common reason for this?
   - □ A buildup of mucus in the kidneys causes damage
   - □ Some medications prescribed to treat CF can damage the kidneys
   - □ Excess salt in the body can damage the kidneys
   - □ Infection in the lungs can spread to the kidneys and damage them
   - □ I’m not sure

7. Can bone fractures be associated with CF?
   - □ Yes
   - □ No
CF Knowledge - Lung and GI topics

8. Why do people with CF often have more lung infections that people without CF?
   - People with CF have more allergies than people without CF
   - People with CF have weak immune systems that cannot fight infection
   - People with CF have mucus in their lungs that easily traps bacteria
   - People with CF have malformed or underdeveloped lungs
   - I’m not sure

9. Does every person with two CF gene mutations develop lung problems?
   - Yes
   - No
   - I’m not sure

10. Should people with CF who have a persistent cough stay at home from work or school to avoid infecting their peers?
    - Yes
    - No
    - I’m not sure

11. When is physiotherapy (to clear the lungs of mucus recommended)
    - Only when a person with CF is feeling sick
    - Only when a person with CF is feeling well
    - All the time, regardless of whether a person with CF is sick or well
    - I’m not sure

12. Are people with CF discouraged from exercising and participating in a sport because it might make them cough?
    - Yes
    - No
    - I’m not sure

13. Does every person with two CF gene mutations develop gastrointestinal (involving the stomach and intestines) problems?
    - Yes
    - No
    - I’m not sure
14. Which organ (often affected by CF) produces and releases defective enzymes?
   - The pancreas
   - The stomach
   - The kidney
   - The intestine
   - I’m not sure

15. During digestion, what is the MAIN nutrient that people with CF have trouble absorbing?
   - Salt
   - Calcium
   - Fat
   - Iron
   - I’m not sure

16. What meal below requires more enzymes than any other to help with digesting food?
   - Chicken sandwich
   - Lasagna and chips
   - Roast chicken, boiled potatoes, and vegetables
   - Chips and gummy candy
   - I’m not sure

17. Which of the following is a sign of food malabsorption (when the body has trouble digesting food)? (Check all that apply)
   - Increased lung infections
   - Excess coughing
   - Greasy stools
   - Clubbing of the fingers
   - Abdominal pain
   - Bloating/swelling of the abdomen
   - Nasal polyps
   - I’m not sure

**CF Knowledge - Reproduction and genetics**

18. Does CF affect male fertility?
   - Yes, most males with CF are infertile
   - Yes, some males with CF have reduced fertility
19. Does CF affect female fertility?
□ Yes, most females with CF are infertile
□ Yes, some females with CF have reduced fertility
□ No, it does not affect female fertility at all
□ I’m not sure

20. Is contraception (to prevent pregnancy) necessary for females with CF who do not wish to have children?
□ Yes
□ No
□ I’m not sure

21. Does a CF carrier always pass the CF gene mutation on to all of his/her children?
□ Yes
□ No
□ I’m not sure

22. If both parents are carriers of CF, what is the chance of having a child who has CF?
□ 0 (0%)
□ 1 in 2 (50%)
□ 1 in 4 (25%)
□ 1 in 10 (10%)
□ 1 in 100 (1%)
□ The child will definitely have CF (100%)
□ I’m not sure

23. IF both parents are carriers of CF, what is the chance of having a child who is neither affected nor a carrier?
□ 0 (0%)
□ 1 in 2 (50%)
□ 1 in 4 (25%)
□ 1 in 10 (10%)
□ 1 in 100 (1%)
□ The child will definitely not be affected nor be a CF carrier
□ I’m not sure
24. If both parents are carriers of CF, what is the chance of having a child who is a carrier?

- [ ] 0 (0%)
- [ ] 1 in 2 (50%)
- [ ] 1 in 4 (25%)
- [ ] 1 in 10 (10%)
- [ ] 1 in 100 (1%)
- [ ] The child will definitely be a CF carrier (100%)
- [ ] I’m not sure

25. If one parent has CF and the other is a carrier for CF, what is the chance of having a child who has CF?

- [ ] 0 (0%)
- [ ] 1 in 2 (50%)
- [ ] 1 in 4 (25%)
- [ ] 1 in 10 (10%)
- [ ] 1 in 100 (1%)
- [ ] The child will definitely have CF (100%)
- [ ] I’m not sure

26. Approximately how many people in America carry one CF gene mutation (what is the carrier frequency?)

- [ ] 1 in 10
- [ ] 1 in 30
- [ ] 1 in 50
- [ ] 1 in 100
- [ ] I’m not sure
APPENDIX H

Self Management Behaviors Questionnaire
In the past 3 months how have you watched for lower respiratory infection in your child. Check (✓) how often you have done each action

<table>
<thead>
<tr>
<th></th>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Usually</th>
<th>Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Observed how fast breathing at rest as a sign of a respiratory infection?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Watched for changes in activity (energy level) in exercise, play, or work?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Watched for changes in hunger/appetite as a sign of a respiratory infection?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Watched for changes in mood/temper?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Watched for weight loss as a sign of a respiratory infection?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Watched for decreased food intake as a sign of respiratory infection?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Watched for increased cough?</td>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8. Watched for increased sputum (mucus)?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>9. Watched for change in the color or thickness of mucus as a sign of a respiratory infection?</td>
<td></td>
<td></td>
<td></td>
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</tr>
</tbody>
</table>
Think about how you treat a lower respiratory infection in your child. Check (√) how often you do each action. NOTE: if your child has not had a lower respiratory infection skip to question 18.

<table>
<thead>
<tr>
<th></th>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Usually</th>
<th>Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>10. Begin respiratory therapy (aerosol, inhalation) or increase the number of respiratory treatments done each day?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>11. Begin chest physical therapy (CPT) or increase the number of CPT sessions done each day?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>12. Begin prescribed antibiotics or increase number of antibiotics used with the advice of your doctor’s office?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>13. Arrange daily time schedule to fit in extra therapy?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>14. Contact the doctor’s office if no improvement occurs after change in therapy?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>15. Stopped treatment at the end of a prescribed course if the symptoms have not disappeared?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
</tbody>
</table>
**Thing about the last time you treated your child for a lower respiratory infection. Check (✓) how often you did each action. Note: if your child has not had a lower respiratory infection skip to question 18.**

<table>
<thead>
<tr>
<th>Question</th>
<th>1-3 days</th>
<th>4-6 days</th>
<th>7-13 days</th>
<th>2-4 weeks</th>
<th>Greater than a month</th>
</tr>
</thead>
<tbody>
<tr>
<td>16. How long did you observe respiratory symptoms before treatment?</td>
<td>□</td>
<td>□</td>
<td>□</td>
<td>□</td>
<td>□</td>
</tr>
<tr>
<td>17. How long did you wait after beginning treatment to decide if there was improvement?</td>
<td>□</td>
<td>□</td>
<td>□</td>
<td>□</td>
<td>□</td>
</tr>
</tbody>
</table>
Think about the *last week* your child used CPT (includes vest). Check (✓) how often you did each action. Note: Skip to 23 if CPT has not been prescribed for your child.

<table>
<thead>
<tr>
<th></th>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Always</th>
<th>Did not Occur</th>
</tr>
</thead>
<tbody>
<tr>
<td>18. Encouraged child to cough intentionally to bring up mucus?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>19. Observed the amount of mucus to decide of CPT was effective?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>20. Observed cough to decide of CPT was effective?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>21. When unable to do CPT with your child made arrangements for someone else to do it?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
<tr>
<td>22. Missed how often CPT sessions were missed during the last week?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
<td>☐</td>
</tr>
</tbody>
</table>

None | Almost none | 1-4 | 1-2 | 4 or more |
Think about the *last week* your child used respiratory therapy (RT)? Note: if no RT prescribed skip to question 34.

<table>
<thead>
<tr>
<th>Question</th>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Always</th>
<th>Did not Occur</th>
</tr>
</thead>
<tbody>
<tr>
<td>23. Observed cough to decide of RT was effective?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>24. Observed mucus production to decide of RT was effective?</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>25. Increased the number of RT treatments with an increase in symptoms?</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>26. Missed how many respiratory treatments during the last week used?</td>
<td>Not Prescribed</td>
<td>None</td>
<td>Almost none</td>
<td>1-4</td>
<td>1-2</td>
</tr>
</tbody>
</table>
For the 3 months, how have you watched digestion problems (malabsorption) in your child? Note: if your child does not have malabsorption (pancreatic insufficiency) skip to question 38.

34. Adjust dose of pancreatic enzymes depending on the amount of food eaten?

35. Adjust does of pancreatic enzymes depending on the type of food eaten (fruit, meat, dairy)?

36. Give enzymes before or during meals or snacks?

37. Make enzymes available to use with meals or snacks eaten away from home?
Think about your child’s diet and eating behavior over the past 3 months. Check (✓) how often you did each action.

<table>
<thead>
<tr>
<th></th>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Usually</th>
<th>Always</th>
</tr>
</thead>
<tbody>
<tr>
<td>38. Observed food intake?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>39. Observed body appearance for signs of weight loss?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>40. Noted fit of clothes as a sign of weigh gain or loss?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>41. Went over growth/weight trend (growth chart) with a member of the health care team?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>42. Used high calorie snacks to add extra calories to diet?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>43. Added more salt diet for increased sweating (as in warm weather)?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>44. Increased calorie intake when weight loss or poor growth was noticed?</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
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<td>☐</td>
</tr>
</tbody>
</table>
45. Discussed poor weight gain with a member of the health care team?

<table>
<thead>
<tr>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Usually</th>
<th>Always</th>
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</thead>
<tbody>
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</tbody>
</table>

46. Missed home many doses of vitamins in the past week?

<table>
<thead>
<tr>
<th>Never</th>
<th>Rarely</th>
<th>Sometimes</th>
<th>Usually</th>
<th>Always</th>
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</table>
APPENDIX I

Research Subject Information and Consent Form
RESEARCH SUBJECT INFORMATION AND CONSENT FORM

TITLE: Influence of Contextual Factors and Self Efficacy on Self Management in Parents of Children with Cystic Fibrosis

VCU IRB NO.: HM20004830

If any information contained in this consent form is not clear, please ask the study staff to explain any information that you do not fully understand. You may take home an unsigned copy of this consent form to think about or discuss with family or friends before making your decision.

PURPOSE OF THE STUDY

The purpose of this research study is to find out about your general knowledge of Cystic Fibrosis, confidence in handling your child’s health needs, ability to manage their care, and the complexity of the medications and treatment they need.

You are being asked to participate in this study because you have a child that has Cystic Fibrosis (CF).

DESCRIPTION OF THE STUDY AND YOUR [YOUR CHILD’S] INVOLVEMENT

If you decide to be in this research study, you will be asked to sign this consent form after you have had all your questions answered and understand what will happen to you.

Participation in this study is a one time commitment. In this study you will be asked to complete a series of questionnaires. Completing the questionnaires will take approximately 20-25 minutes. After you complete the questionnaires, your child’s medical record will be reviewed to obtain information about medications and treatments they are taking and results of their pulmonary function test (PFT’s) if available.

RISKS AND DISCOMFORTS

Sometimes people get upset about the time that it will take you to complete the survey. We anticipate that it will take 20-25 minutes and you will be able to take breaks as necessary.

USE AND DISCLOSURE OF PROTECTED HEALTH INFORMATION
Authority to Request Protected Health Information

The following people and/or groups may request my Protected Health Information:
- Principal Investigator and Research Staff
- Research Collaborators
- Data Safety Monitoring Boards
- Others as Required by Law
- Study Sponsor
- Institutional Review Boards
- Government/Health Agencies

Authority to Release Protected Health Information

The VCU Health System (VCUHS) may release the information identified in this authorization from my medical records and provide this information to:
- Health Care Providers at the VCUHS
- Study Sponsor
- Data Coordinators
- Data Safety Monitoring Boards
- Others as Required by Law
- Principal Investigator and Research Staff
- Research Collaborators
- Institutional Review Boards
- Government/Health Agencies

Once your health information has been disclosed to anyone outside of this study, the information may no longer be protected under this authorization.

Type of Information that may be Released

The following types of information may be used for the conduct of this research:
- Complete health record
- Diagnosis & treatment codes
- Discharge summary
- History and physical exam
- Consultation reports
- Progress notes
- Laboratory test results
- X-ray reports
- X-ray films / images
- Photographs, videotapes
- Complete billing record
- Itemized bill
- Information about drug or alcohol abuse
- Information about Hepatitis B or C tests
- Information about psychiatric care
- Information about sexually transmitted diseases
- Other (specify):

Expiration of This Authorization

This authorization will expire when the research study is closed, or there is no need to review, analyze and consider the data generated by the research project, whichever is later.
☐ This research study involves the use of a Data or Tissue Repository (bank) and will never expire.
☐ Other (specify):

Right to Revoke Authorization and Re-disclosure

You may change your mind and revoke (take back) the right to use your protected health information at any time. Even if you revoke this Authorization, the researchers may still use or disclose health information they have already collected about you for this study. If you revoke this Authorization you may no longer be allowed to participate in the research study. To revoke this Authorization, you must write to the Principal Investigator.

BENEFITS TO YOU AND OTHERS

You may not get any direct benefit from this study, but, the information we learn from people in this study may help us design better programs to support families who have children with CF.

COSTS

There are no costs for participating in this study other than the time you will spend filling out the questionnaires.

Alternatives

Your alternative is not to participate in this study.

CONFIDENTIALITY

Potentially identifiable information about you will consist of questionnaires and information taken from the medical record. Data is being collected only for research purposes.

Your data will be identified by ID numbers not names, and stored separately from research data in a locked research area. All personal identifying information will be kept in password protected files and these files will be deleted after 5 years. Other records such as completed questionnaires will be kept in a locked file cabinet for 5 years after the study ends and will be destroyed at that time. Access to all data will be limited to study personnel. A data and safety monitoring plan is established.

We will not tell anyone the answers you give us; however, information from the study and information from your child’s medical record and the consent form signed by you may be looked at or copied for research or legal purposes, or by Virginia Commonwealth University. Personal information about you might be shared with or copied by authorized officials of the Department of Health and Human Services or other federal regulatory bodies.
What we find from this study may be presented at meetings or published in papers, but your name will not ever be used in these presentations or papers.

**VOLUNTARY PARTICIPATION AND WITHDRAWAL**
You do not have to participate in this study. If you choose to participate, you may stop at any time without any penalty. You may also choose not to answer particular questions that are asked in the study.

Should you decide you wish to withdraw your date you can notify the PI or student investigator by either written or oral communication.

Your participation in this study may be stopped at any time by the study staff without your consent. The reasons might include:

- the study staff thinks it necessary for your health or safety;
- you have not followed study instructions;
- the sponsor has stopped the study; or
- administrative reasons require your withdrawal.

**QUESTIONS**
If you have any questions, complaints, or concerns about your participation in this research, contact:

Jeanne Salyer Ph.D., RN  
School of Nursing  
Virginia Commonwealth University  
1100 East Leigh Street  
Richmond, VA 23239  
Telephone: (804) 828-2773

Erin Booth, MS, RN  
Telephone: (804) 516-4925

The researchers/study staff named above are the best person(s) to call for questions about your participation in this study.

If you have any general questions about your rights as a participant in this or any other research, you may contact:

Office of Research  
Virginia Commonwealth University  
800 East Leigh Street, Suite 3000
Contact this number to ask general questions, to obtain information or offer input, and to express concerns or complaints about research. You may also call this number if you cannot reach the research team or if you wish to talk with someone else. General information about participation in research studies can also be found at http://www.research.vcu.edu/irb/volunteers.htm.

CONSENT
I have been given the chance to read this consent form. I understand the information about this study. Questions that I wanted to ask about the study have been answered. My signature says that I am willing to participate in this study. I will receive a copy of the consent form once I have agreed to participate.

Participant name printed          Participant signature          Date

___________________________________________________________________________
Name of Person Conducting Informed Consent
Discussion / Witness
(Printed)

___________________________________________________________________________
Signature of Person Conducting Informed Consent          Date
Discussion / Witness
Principal Investigator Signature (if different from above)    Date
Erin Booth was born on September 15, 1982, in Richmond, Virginia. She graduated from James River High School, Midlothian, Virginia in 2000 and received her Bachelor of Science in Nursing from East Carolina University, Greenville, North Carolina in 2004. In 2008 she received her Master of Science from Virginia Commonwealth University with a focus in Leadership and Administration. Throughout her career she has been employed in a variety of settings including inpatient, outpatient, and pharmacy care of individuals with Cystic Fibrosis.