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Abstract

Historically, individuals living with cystic fibrosis (CF) gathered strength and support attending summer camps, support groups, and community events. However, in 2003, medical guidelines recommended stringent separation of individuals with CF to reduce the risk of cross-infection and disease progression. This study seeks to examine the impact of these recommendations on the social experiences of young adults with CF. A qualitative, exploratory, grounded theory approach was employed to understand the experience of young adults living with CF, with specific emphasis on the impact of *social connection and isolation* on their lives. Semi-structured individual interviews were conducted with ten young adults aged 18 – 25 diagnosed with CF. An interview guide was used to elicit information about how individuals with CF understand, experience, and perceive contact and social isolation from others with CF, ways in which they experience their social worlds, and the impact isolation from others with CF has on their medical and psychosocial development during the young adult years. Interviews were digitally recorded, transcribed verbatim, and coded using grounded theory's constant comparative method. Analysis identified the importance of *social connection and isolation* as a common theme, supported by young adults' use of social media, familiarity with the infection prevention and control guidelines, relationships with their siblings, the value of their friendships, and supported autonomy. Participants struggled to *define normal* in their daily lives. Increased social work practitioners' awareness of ways to support young adults with chronic illness to engage with the CF community may improve health and mental health outcomes.

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“I am not CF, I have CF”: Social Connection and Isolation During Young Adulthood With

Cystic Fibrosis

Chelsea Elizabeth Toth

A DISSERTATION

In

Social Work

Presented to the Faculties of the University of Pennsylvania

In

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Degree of Doctor of Social Work

2016

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Dedication

This dissertation is dedicated in loving memory to CF warrior Korinna Conron.

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ABSTRACT

“I AM NOT CF, I HAVE CF”: SOCIAL CONNECTION
AND ISOLATION DURING YOUNG ADULTHOOD WITH CYSTIC FIBROSIS

Chelsea Elizabeth Toth

Allison Werner-Lin, PhD

Historically, individuals living with cystic fibrosis (CF) gathered strength and support attending summer camps, support groups, and community events. However, in 2003, medical guidelines recommended stringent separation of individuals with CF to reduce the risk of cross-infection and disease progression. This study seeks to examine the impact of these recommendations on the social experiences of young adults with CF. A qualitative, exploratory, grounded theory approach was employed to understand the experience of young adults living with CF, with specific emphasis on the impact of *social connection and isolation* on their lives. Semi-structured individual interviews were conducted with ten young adults aged 18 – 25 diagnosed with CF. An interview guide was used to elicit information about how individuals with CF understand, experience, and perceive contact and social isolation from others with CF, ways in which they experience their social worlds, and the impact isolation from others with CF has on their medical and psychosocial development during the young adult years. Interviews were digitally recorded, transcribed verbatim, and coded using grounded theory’s constant comparative method. Analysis identified the importance of *social connection and isolation* as a common theme, supported by young adults’ use of social media, familiarity with the infection prevention and control guidelines, relationships with their siblings, the value of their friendships, and supported autonomy. Participants struggled to *define normal* in their daily lives. Increased social work practitioners’ awareness of ways to support young adults with chronic illness to engage with the CF community may improve health and mental health outcomes.

Keywords: cystic fibrosis, social connection, social isolation, chronic illness, young adulthood, qualitative research, grounded theory

Chapter 1

Introduction

Individuals living with cystic fibrosis (CF) represent a unique patient population because most acquire difficult to treat pathogens in their respiratory tract. Epidemiologists have studied the impact of these pathogens, risk factors and rates of transmission to individuals living with CF. Infection control recommendations for patients with cystic fibrosis (2003) advised separating individuals living with CF by a minimum distance of three feet from others diagnosed with CF. Ten years later, updated guidelines presented additional research related to the transmission of pathogens, and subsequently increased the distance between individuals with CF to six feet (Saiman et al, 2014).

Cystic fibrosis (CF) is an inherited chronic disease that is unique for a number of reasons. The prescribed daily treatment regimen is often complex, including multiple airway clearance sessions and large numbers of oral and inhaled medications. In addition to this high treatment burden, patients suffer from chronic respiratory tract infections that have led to the development of strict infection-control measures in the outpatient and inpatient medical care environments. The theoretical benefit of these isolation practices is to prevent infection, spread from one CF individual to another, but these practices also frequently result in social isolation which can have profound effects on an individual's emotional coping and adherence behaviors. The effects of social isolation among young adult patients with cystic fibrosis are unclear; often this creates a major adherence barrier and impairs the achievement of successful treatment, and can ultimately affect long-term morbidity and mortality.

There remains a need to better understand the experience and scope of social isolation among young adults with CF and to describe its perception and impact on the transition from adolescence to adulthood. Increased understanding of social isolation may permit development

of specific interventions to help young adult CF patients identify and reduce their social isolation, to achieve better long-term health and quality of life.

Young adults with cystic fibrosis account for more than half of all individuals living with cystic fibrosis (CF) within the United States (Cystic Fibrosis Foundation Patient Registry, 2014). Historically, CF was a childhood disease, today individuals with this chronic illness are surpassing expectations living beyond early adulthood. The purpose of this research is to explore the current gap in the literature examining the impact of social isolation related to disease management, disease progression and life expectancy. This traditional book style, qualitative dissertation research will describe the impact of social isolation among young adults living with cystic fibrosis.

CHAPTER 2

Review of the Literature

Research exploring psychosocial impacts of cystic fibrosis on the individuals living with the disease is still developing and currently limited. The literature review will provide an overview of information related to the purpose of this study. The literature will allow the reader to deepen their understanding of the disease process, medical treatments, challenges of treatment adherence, contact precautions and isolation, psychosocial impact, anxiety and depression, and young adulthood while identifying the gaps in existing research.

Disease Process in Cystic Fibrosis

Cystic fibrosis is the most common life-shortening genetic disease in the Caucasian population (Jennings, Riekert, & Boyle, 2014) affecting 1 in 3,500 individuals (Cystic Fibrosis Foundation Patient Registry, 2012). It is an autosomal recessive disorder that is usually diagnosed in early infancy based on the results of newborn screening (Jennings et al., 2014; Nufeld & Keith, 2012). More specifically cystic fibrosis is a

... genetic disorder that affects the respiratory, digestive and reproductive systems involving the production of abnormally thick mucus linings in the lungs and can lead to fatal lung infections. The disease can also result in various obstructions of the pancreas, hindering digestion (World Health Organization, 2012, p.5).

Newborn screening is routinely performed in the United States, Australia and numerous European countries (Zemanick et al., 2010). If the newborn screening is positive, the patient is subsequently evaluated with a sweat chloride test and possibly genetic testing to confirm the diagnosis of CF (Cystic Fibrosis Foundation, 2014).

The prevalence of cystic fibrosis is about one in every 3,500 births in the United States, and one in every 3,600 births in Canada (Cystic Fibrosis Foundation Patient Registry, 2012; Cystic Fibrosis Canada, 2014). These numbers are lower in comparison to one in every 2,000-

3,000 births in the European Union (World Health Organization, 2012). In Canada there are roughly 4,000 individuals living with CF (Cystic Fibrosis Canada, 2014). In the United States there are approximately 30,000 individuals living with CF and 70,000 worldwide (Cystic Fibrosis Foundation Patient Registry, 2012).

Individuals diagnosed with CF have a greater life expectancy today than ever before, as a result of major advances in diagnosis and treatment (Szyndler, Towns, Asperen & McKay, 2005). Once known as a disease of childhood, individuals with CF are now living into adulthood. Between 1969 and 1990, the average age of individuals living with CF doubled, from 14 to 28 years (Elgudin, Kishan, & Howe, 2004). In 2014, the average age of adults living with CF was 39 years (Cystic Fibrosis Foundation Patient Registry, 2014). Over half the population of individuals living with CF in the United States are older than 18 years and the oldest person living with cystic fibrosis on record is 86 years old (Cystic Fibrosis Foundation Patient Registry, 2014). Longer life expectancy increases the need to understand the experience of individuals with CF who age out of pediatric medical practices, to better navigate the developmental tasks of adulthood.

Medical Treatment

Although there is currently no cure for cystic fibrosis (Cystic Fibrosis Canada, 2014), early diagnosis permits early institution of preventative therapies and leads to improved outcomes (Ernst, Johnson, & Stark, 2010). Individuals with CF often have respiratory, sinus, and digestive comorbidities. Following diagnosis, the treatment regimen usually includes therapies such as inhaled medications, oral medications, intermittent intravenous (IV) antibiotics, regular airway clearance, nutritional supplements, vitamins, and pancreatic enzyme replacements (Elgudin et al., 2004; Quittner et al., 2012). Prescribed airway clearance regimens include twice daily airway clearance therapies when healthy, increasing to four times a day during respiratory illness (Quittner et al., 2012; Abbott & Gee, 2003). Airway clearance should be performed even

when an individual with CF is feeling well to maintain lung function and overall health (Cystic Fibrosis Foundation Patient Registry, 2012); each session generally takes 20-30 minutes to complete.

Cystic fibrosis care is centralized to accredited CF centers across the United States and they utilize a multidisciplinary medical team approach to care (Tuchman, Schwartz, Sawicki, & Britto, 2010). In 2014, over 50% of individuals with CF in the United States were over the age of 18 years, approximately 29% more than reported in 1986 (Cystic Fibrosis Foundation Patient Registry, 2014). In response to the growing number of adult patients, the Cystic Fibrosis Foundation developed a policy mandating all CF centers that have more than 40 adult patients establish and accredit an adult CF center (Tuchman et al., 2010). Many centers share medical staff and utilize the model “1 center, 2 programs” (Tuchman et al. 2010, p. 568).

Most CF specialists recommend regular follow up appointments at least four times a year (Cystic Fibrosis Foundation Patient Registry, 2014; Nuefeld & Keith, 2012). The CF medical teams remain focused on reducing cross infection for patients in their practice. Medical teams aim to develop individualized treatment plans and regimens in consensus with patients and their families to best accommodate the patient’s daily life. The medical team’s primary goal is to prevent structural lung changes that inevitably develop from chronic and recurrent lung infections and inflammation. Respiratory infections require frequent antibiotics which can be given as either oral, inhaled or intravenous depending on the disease severity (Abbott & Gee, 2003; Nuefeld & Keith, 2012). CF patients may face dangerous repercussions if they neglect to follow their prescribed treatments (Ernst et al., 2010). If the patient does not significantly respond to outpatient antibiotic therapy, he or she will often be admitted to the hospital for more aggressive medical treatment (Neufeld & Keith, 2012).

Inpatient hospitalization for cystic fibrosis treatment provides an environment that has the ability to encompass a multi-system approach and includes IV antibiotics and aggressive airway

clearance. In the hospital setting, patients interact with providers who can help them to understand the disease, improve health outcomes, discuss compliance, change lifestyle and shift treatment regimes to improve patient quality of life (Jung, Heinrichs, Geidel, & Lauener, 2012). The first hospital admission in an adult-oriented hospital setting can be a jarring experience. The initial hospitalization in an adult-oriented centered CF center is often challenging for both the patient and the medical team. Early on, there may be feelings of trepidation among patients as their new medical team becomes fully acquainted with that individual's history and care needs.

Challenges of Treatment Adherence

Early diagnosis, effective therapies and a multidisciplinary medical team approach has led to improved health outcomes and have extended the life expectancy of individuals with CF. Despite these improvements, CF remains a progressive and life-shortening multi-system disease (Abbott & Gee, 2003; Modi & Quittner, 2003). According to Ernst et al. (2010), 95% of patients with CF will die from pulmonary complications. Lack of adherence with recommended treatments and contact restrictions may lead to increased frequency of respiratory exacerbations and infection, resulting in decreased pulmonary function, increased hospitalizations, and shorter life span (Abbott and Gee, 2003; Modi & Quittner, 2006; Nuefeld & Keith, 2012). "Poor adherence has been cited as the single greatest cause of treatment failure" (Quittner, Modi, Lemanek, Ievers-Landis, & Rapoff, 2008, p. 917). The ramifications of poor treatment adherence include increasing morbidity and mortality, as well as reduction in an individual's quality of life (Modi & Quittner, 2006; Quittner et al., 2008).

Individuals with CF are aware of the shortened life expectancy associated with their disease, and this knowledge imposes a challenge on morbidity and mortality factors (Modi & Quittner, 2006; Quittner et al., 2008). This awareness predisposes to many behavioral, emotional and cognitive challenges for individuals with CF and their families (Ernst et al., 2010; Zemanick et al., 2010). For example, individuals with CF often participate in high-risk behavior due to the

knowledge of their shortened life expectancy, or wait until their disease is uncontrolled to obtain medical care. A comprehensive approach to care – including patient and family education, behavior modification and opportunities for social support – is essential to improving self-management (Jennings et al., 2014) and reducing disease complications and mortality. Factors associated with improved adherence include: attending routine appointments at CF specialty clinics, obtaining the results from pulmonary function test, demonstrated increase in BMI, and perceived health benefits and social support (Jennings et al., 2014).

Contact Precautions and Isolation

“Over the past 20 years there has been a greater interest in infection control in cystic fibrosis (CF) as patient-to-patient transmission of pathogens has been increasingly demonstrated in this unique patient population” (Saiman & Siegel, 2004, p.57). Despite continuous adherence to prescribed treatments, individuals with CF have complex, difficult to treat pathogens in their respiratory tract. The most common infections include, *Staphylococcus aureus* and *Pseudomonas aeruginosa* (Saiman, Seigel, & the Cystic Fibrosis Foundation Consensus Conference on Infection Control Participants, 2003). In addition to individual’s respiratory infection, the prevention of cross infection among patients is a major concern in the CF community. Cross infection occurs between individuals with CF when respiratory virus-causing pathogens and other infections are transmitted through air particles (Saiman & Siegel, 2004; Saiman et al, 2014).

Risks of shared infection among people with CF shapes the care and daily social lives of patients in many ways. Pathogens increase incidence of new infections in some patients, which may require treatment with increased antibiotics. Unnecessarily acquiring a respiratory pathogen from another CF patient may result in more dangerous infection and decrease a patient’s quality of life and impact their survival. To reduce the risk of dangerous cross infection, medical teams must follow contact precautions while working with CF patients including wearing a mask,

gown, and gloves throughout any clinical contact (Nuefeld & Keith, 2012; Saiman et al., 2014).

The guidelines advise medical teams to educate patients and their families on isolation precautions to prevent patient-to-patient transmission of respiratory pathogens.

Since the publication of *Infection Control in Cystic Fibrosis* in 2004, there have been advances in the practice of infection control and prevention in cystic fibrosis (Saiman et al., 2014). Due to the documented risk of patient-to-patient transmission of pathogens, the updated *Infection Prevention and Control Guideline for Cystic Fibrosis* prescribes the necessity to “separate all people with CF from others with CF, regardless of their respiratory tract culture results, at least 6 feet (2 meters) in all settings, to reduce the risk of droplet transmission of CF pathogens” (Saiman et al., 2014, p. S4). Contrastingly, previous infection control guidelines that stated that an individual with cystic fibrosis should not be within 3 feet of another person with CF due to the risk of cross infection (Saiman et al., 2003).

The infection prevention and control guidelines encourage CF health care teams to educate patients and their families about contact precautions (Saiman et al., 2014). Having recurring conversations with the patient and their family about the importance of adherence with the barriers to care can normalize the difficulties these families endure (Modi et al., 2006). The stringent guidelines can, however, lead to psychosocial complications, including feelings of isolation (Saiman et al., 2003, Saiman et al., 2014). Medical teams are recommended to encourage the patient and their family to express their concerns regarding the implications of contact precautions.

For many other chronic diseases, social support is essential in promoting adherence while validating and normalizing patient experiences. This support in the CF community, however, is limited by the dangers associated with the contact from others with CF. Due to frequent illnesses and multiple hospitalizations, adolescents with chronic illness may have less social interactions than their peers, which increases their risk of impaired emotional development (Johnson, Ravert,

& Everton, 2001). An obstacle specific to individuals with CF is the isolation from other patients with CF (Johnson et al., 2001; Saiman et al., 2014). Historically, individuals with cystic fibrosis would come together for support through summer camps. These camps included a high-calorie diet, chest physiotherapy, and daily physical activities, which improved overall health outcomes (Blau et al., 2002). Today these CF camps and support groups are relics of the past, as studies of the camps revealed that patient to patient contact increased the risk of cross infection. In response to these findings, in 1993 the U.S. Cystic Fibrosis Foundation issued a recommendation to discontinue CF summer camps (Pegues et al., 1994; Blau et al., 2002). Instead, CF patients were encouraged to participate in activities including sports and camps with individuals not diagnosed with CF.

The encouragement to participate in activities with non-CF individuals allows patients to socialize, but patients with CF subsequently lack connection with like peers. Individuals with CF do not have the accessibility to connect with like peers face to face who understands the daily CF lifestyle. The protection offered by these guidelines is important for prevention of cross infection, but the disconnect impacts individuals living with CF, isolating them from others with CF.

Psychosocial Impact of Cystic Fibrosis

A new era for cystic fibrosis care has emerged as the first generation of patients survive and grow into adulthood. Young adulthood fosters unique challenges in future life plans, including career goals, finding a partner, and family planning. Consequences from the choices made during adolescence, including treatment adherence, likely have an impact on adulthood (Tuchman et al, 2010). Patients with CF are required to acclimate to practice changes associated with the advances in medical care and therapies. As such, updated treatment guidelines require enforcing stricter isolation precaution protocols today than in the past (Saiman et al., 2014). The

development of subsequent social isolation has multiple implications and ramifications for the daily life of those living with CF.

Psychological and family functioning are important factors for those involved in the daily care of individuals with cystic fibrosis (Szyndler et al., 2005). Often isolated from their peers due to worsening disease and increased admissions to the hospital, CF patients frequently rely on their family for support (Ernst et al., 2010). Adults with CF often lack the motivation to engage in social activities due to anxiety, thus impeding interpersonal relationships (Yohannes, Willgoss, Fatoye, Dodd, & Webb, 2012). Peer relationships are important for young adults as they transition to independence and self-management, offering elements of socialization that are often beneficial to those suffering with chronic illness (Johnson et al., 2001). Nonetheless, due to the strict infection control guidelines, there are no CF-specific face to face groups or activities creating a paucity of interpersonal emotional support.

While there are many barriers that limit how CF patients can socialize with their CF peers, advances in technology afford patients with opportunities to communicate more readily with each other. For example, individuals with cystic fibrosis may communicate through the internet with other CF patients (Saiman et al, 2014), while the use of smart phones and wireless internet connections provide accessibility to peers and medical information. In 2001, Hopkins Teen Central was launched as an online support group for adolescent patients with CF (Johnson et al., 2001). Electronic equipment was provided to those participants in need. This innovative online support group provided adolescents living with CF the opportunity to discuss issues at the nexus of CF and typical adolescence (Johnson et al., 2001). Technology increases the opportunity for those living with CF to connect with each other via text message or the internet, without the risk of cross infection (Cystic Fibrosis Foundation Patient Registry, 2012).

Anxiety and Depression in Cystic Fibrosis

Anxiety and depression are common in adult patients with CF and are often undiagnosed (Yohannes et al., 2012). The quality of life for individuals with cystic fibrosis is directly related with their emotional well-being and mental health (Szyndler et al., 2005). It has been demonstrated that an increased illness burden, poor adherence level, and depression are associated with the intensity and complexity of prescribed treatments (Modi & Quittner, 2006), all of which may be intensified due to social isolation from CF peers. Implications of depression and anxiety have also been associated with impaired interpersonal relationships and quality of life, severity of respiratory symptoms, poor lung function, and re-hospitalizations (Yohannes et al., 2012).

Patients with CF who perceive themselves as having a positive future, despite the grim prognosis of their disease, demonstrate better psychological functioning (Szyndler et al., 2005). Conversely, the literature demonstrates that patients living with CF have a high risk of comorbid mental health conditions based on psychosocial factors including the ability to cope with stress and emotional regulation (Ernst et al., 2010). Negative impact is shown when individuals with CF perceive themselves as different from their peers or keep their disease a secret (Ernst et al., 2010). Furthermore, several studies demonstrate poor lung function is associated with elevated levels of depression (Duff et al., 2014).

Several tools have been developed to screen adults with CF routinely for anxiety and depression symptoms (Yohannes et al., 2012). The International Depression/anxiety Epidemiology Study (TIDES) evaluated the prevalence and impact of anxiety and depression for individuals with CF in the United States and eight other countries (Duff et al., 2014). This study utilized the Hospital Anxiety and Depression Scale to evaluate the prevalence incorporating CF specific demographics (Duff et al., 2014). Identifying the needs of the patients from a

comprehensive multidisciplinary approach can lead to improved psychosocial factors (Jung et al., 2012).

Young Adulthood

The level of treatment adherence during young adulthood is often related to the complexities and challenges in the patient's home environment. Common barriers to treatment include time constraints, treatment burden, financial challenges and lack of disease understanding (Zemanick et al., 2010). The time necessary to perform daily treatments can impair a young adult's ability to be active participants with their peers. A balance is needed to achieve satisfactory treatment adherence along with daily activities and peer relationships.

Written treatment plans can assist patients to improve treatment adherence by establishing identifiable goals. A young adult's habits and behaviors begun in adolescence, including avoidance, can often affect adherence behaviors and long term outcomes. Working collaboratively with young adults living with cystic fibrosis to develop mutually agreeable treatment plan, positive reinforcements and consequences, may have beneficial outcomes to their health and overall wellbeing. For example, to increase adherence, medical teams and young adults may identify barriers to adherence and work towards an agreed upon reward for completing the prescribed therapies.

Young adulthood, for individuals with CF, involves a process of moving towards independence, which involves many participants: the patient, their family and medical team. Transitioning to adulthood may involve connected autonomy (Doyle & Werner-Lin, 2014), as a chronically ill young adult maintains the relationship with their family, while gaining their independence, understanding their support network is still intact is essential. Simultaneously, there is a transfer of power from the caregiver parent to the patient with regards to disease treatment and management (Doyle & Werner-Lin, 2014).

The growing adult population living with CF will experience new and unknown challenges. Psychosocial implications including social isolation from peers living with CF are under discussed. This population may realize and live with consequences from decisions made during their adolescence with impact on the health status of current young adults.

Multidisciplinary care for adolescents and young adult CF patients must include increasing awareness of their medical care, understanding of the experience of being socially isolated from peers with CF and the psychological implications.

Research Questions

This study aims to explore three primary and related research questions: How do individuals with CF aged 18-25 understand, experience, and perceive social isolation from others with CF? Are there others ways in which young adults with CF experience isolation? How does social isolation impact their medical and psychosocial development during the young adult years?

CHAPTER 3

Methods

Reflexivity Statement

I identify as a social worker with over four years of experience working with the CF population at a Pediatric Cystic Fibrosis Center. I am familiar with the disease progression, complexities of disease management, treatment regimen, and the Infection Prevention and Control Guidelines. Although there is limited research exploring social isolation among the cystic fibrosis population, I was intrigued by this topic and the effect social isolation may have on young adults living with cystic fibrosis. I provide care as a member of a multidisciplinary medical team for children, adolescent and young adult patients with cystic fibrosis encompassing diverse ethnic backgrounds and varying socioeconomic status.

I have attended yearly North American Cystic Fibrosis Conferences which adds to the breadth of knowledge acquired about this particular population. I am an active participant on the North American cystic fibrosis social worker email list serve. Some of the recurring themes on the list serve include recommendations to decrease isolation between areas, to improve connection between individuals living with CF, and discussion of if peer mentoring would be appropriate. Several patients have expressed their frustrations with the infection guidelines while attending clinic appointments. At clinic appointments, some patients described feeling isolated from others living with CF and had a desire to communicate with them in person. This led me to wonder what patients do to feel less isolated and how they connect to others living with CF. This conceptualization of social isolation piqued my interest to further explore the literature related to isolation and CF, and a gap was noted in the literature related to social isolation for individuals living with this unique chronic illness.

I interpret my role as a cystic fibrosis social worker in several ways which include conducting psychosocial assessments, case management, discussing treatment adherence,

providing school accommodations, assisting patients and families to obtain insurance, and providing appropriate resources and referrals. I understand the importance of prescribed medications and airway clearance. I have witnessed the positive aspects and negative repercussions of patient outcomes based on their level of adherence to prescribed treatments. I perceive the delicate balance supporting adherence with treatment regimen and maintaining a non-judgmental therapeutic relationship.

Knowing this information, I was vigilant to recognize my personal biases and experiences. I judiciously developed open-ended questions to explore participants understanding, perception, and experience of living with cystic fibrosis. I was open to participant's responses that confirmed or denied my own anecdotal clinical evidence. During the interviews, I listened attentively and probed for concrete details. Throughout the process, I wrote memos to observe personal preconceptions and reactions to the session. I also wrote memos to document codes and potential themes as they emerged from the data. I closely reviewed the aforementioned memos which aided in monitoring and managing any bias that occurred. The combination of my clinical work and simultaneous data collection allowed me to observe the broader impact of connection and social isolation among the lives of young adults with CF.

In conducting the face-to-face interviews, I utilized several social work skills. I utilized the skills of establishing rapport during the interview which developed a comfortable and safe space for participants to share their shame, vulnerabilities and overall personal experiences. I was comfortable asking the open-ended questions and would probe for additional information. In addition, I observed a recurrent theme that emerged during the first interview and adapted the interview guide to include questions around that topic. I was comfortable with the use of silence, which aided in participants deepening their reflection and responses. I did occasionally ask closed ended questions to clarify concrete information. I improved upon active listening and

refrained from inserting leading questions or comments as the interviews progressed. Self-awareness was key as I avoided inserting personal thoughts and solving responses.

Design and Setting

The study uses a qualitative, exploratory, grounded theory approach to gain deeper understanding of the experiences and perceptions of social isolation for emerging adults (aged 18-25) living with cystic fibrosis.

The study explored three primary and related research questions:

- How do individuals with CF aged 18-25 understand, experience, and perceive social isolation from others with CF?
- Are there others ways in which young adults with CF experience isolation?
- How do emerging adults understand the impact of social isolation on their medical and psychosocial development during the young adult years?

The study received approval from the University of Pennsylvania's Institutional Review Board (Appendix A).

Sample selection criteria.

Inclusion criteria for this study include females and males who are: (1) English-speaking (2) aged 18-25, and (3) diagnosed with cystic fibrosis over two years prior to interview.

Exclusion criteria include: (1) current hospitalization for a cystic fibrosis-related exacerbation, and (2) young adults who have been diagnosed with cystic fibrosis within the past two years. The sample also excludes siblings of participants; only one individual with a CF diagnosis from any nuclear family living with cystic fibrosis will be interviewed.

Recruitment.

The study aimed to recruit approximately 10-15 participants using convenience and snowball methods. Recruitment occurred through the national CF foundation, its affiliates, and online through social media websites. A recruitment flyer with study details was distributed to organizations that serve individuals with CF (Appendix B), including the local Northeastern PA Chapter and the National Cystic Fibrosis Foundation. The Cystic Fibrosis Foundation is a national nonprofit organization that was established in 1955 to serve people in the United States diagnosed with cystic fibrosis. Currently the Foundation serves approximately 30,000 people living with CF, their families and their communities (Cystic Fibrosis Foundation, 2013).

Additionally emails were sent to: the local chapter of the cystic fibrosis foundation (Appendix C) and the North American cystic fibrosis social work email list-serve (Appendix D). Snowball sampling occurred as participants referred other individuals with CF who meet the eligibility criteria.

Consent.

Recruitment materials included a phone number and email for the researcher, inviting interested parties to make contact. The researcher then responded to telephone and email inquiries to ensure eligibility criteria were met. During initial telephone contact and email correspondence the researcher described the identity and role of the researcher. The researcher explained the study, the time involved, confidentiality, planned use of data and that participation in the study was voluntary and participants were free to withdrawal at any time.

Individuals reviewed the information and informed consent form (Appendix E) on site prior to the individual interview or prior to the internet interview, and consented to digital audio recordings of the interview. The study posed minimal risk to individuals participating.

The information the participants shared was kept strictly confidential. The researcher will never use participant names, identifying information or information about where participants live or work in documenting the interview without written permission. Participants will be described by their age or gender when quotations are necessary to documenting or presenting the findings.

Any printed records are stored under lock and key. The records of this study will be kept confidential and are stored on a password-protected server, accessed only by the primary researcher. The interviews were audio recorded, however, names and other identifying information were not used while recording, as to keep the participant's identity confidential. Only the researchers have access to these digital recordings, and all recordings will be erased from devices and deleted from computer files two years after the conclusion of this study. The researcher cannot guarantee total privacy, as records can be opened by court order or produced in response to a subpoena or a request for production of documents.

Data collection.

Interviews occurred with participants face to face in person and using Zoom, a video conferencing software (a technology similar to Skype). Face-to-Face interviews took place in a setting mutually agreed upon by the interviewer and the research subject. All interviews lasted approximately 60 minutes.

Participants completed a demographic questionnaire (Appendix F), which elicited information including age at diagnosis, current educational and employment status, and insurance coverage. A semi-structured interview guide (Appendix G) was developed in collaboration with seasoned qualitative researchers. The guide addresses: experiences of living with cystic fibrosis, awareness of perspectives on infection prevention and control guidelines,

experiences and beliefs about social isolation, future expectations of illness and development.

The primary questions include:

- Living with CF:
 - “Tell me about living with CF. What is a typical day like for you now? Tell me about the ways in which CF has impacted your daily life? Your relationships with family, friends, romantic partnerships? If you were talking to someone who didn’t know anything about CF, how would you describe it? What you want them to know?”
- Infection Prevention and Control Guidelines:
 - “What do you know about the Infection Prevention and Control Guidelines? Are there ways in which the guidelines impact your daily life? Tell me about these. The guidelines suggest limiting face to face contact from other individuals with CF, how does this impact your daily life?”
- Social Isolation:
 - “Do you interact or communicate with other people with have CF? How? Tell me about a situation that made you feel isolated from others with CF? What are some avenues you use to feel less isolated?”

The researcher had a brief conversation with the participant before starting the audio recording, during which time the researcher clarified the purpose, obtained consent, discussed confidentiality and developed initial rapport. The researcher made an audio recording of the interview and took written notes during the interview. Immediately after the interview the researcher wrote memos to record observations and identify potential categories (Charmaz, 2006). Each participant received a \$15 Amazon gift card sent to their preferred email address at the completion of the interview.

Each interview participant received a unique alphanumeric identifier linking individual audio recording and transcript to participants. The interview was transcribed verbatim from the audiotape, using an external transcription service, ADA Transcription – these were the only individuals besides the researcher who listened to the audiotape. The transcription service received the digital recording with the alphanumeric identifier the researcher reviewed the transcript along with the audio recording to ensure accuracy. The researcher removed any spoken information that might serve to identify participants, including geographic locations and names the participant mentioned during the interview. The researcher analyzed the interview and read the transcription for this research.

Methods of Data Analysis, A Grounded Theory Approach

The researcher utilized a grounded theory approach to explore the perception, experience and understanding of young adult participants in this study. Grounded theory provided a systematic approach to analyze the data collected, generate concepts and construct theory (Charmaz, 2006). The study employed qualitative methodology which utilized an inductive, subjective and contextual approach to develop concepts as they emerged from the data (Morgan, 2014).

The researcher's role during the interview was to observe with sensitivity, listen and encourage the participant to respond (Charmaz, 2006). The researcher remained flexible during the semi-structured interviews and understood the importance of the meaning identified by the participants (Morgan, 2014). The open-ended questions on the interview guide allowed the researcher to probe for additional information and pursue topics that emerged during the conversation (Morgan, 2014). To acknowledge personal subjectivity (Morgan, 2014) the researcher wrote memos throughout the analytic process (Charmaz, 2006).

The line-by-line coding process described by Charmaz (2006) is to prompt the researcher to observe details and remain open to the data. Memo writing allowed the researcher to recognize concepts and relationships between codes (Charmaz, 2006). Line-by-line coding allowed the researcher to develop open codes, recognize *in vivo* codes (participant's own words), and identify the content (Charmaz & Bryant, 2011). Through memo writing the researcher identified, clarified and compared codes (Charmaz & Belgrave, 2012). The researcher then condensed and sorted the open codes to develop focused codes to further categorize the data (Charmaz, 2006). The focused coding process is a strength of the grounded theory approach because it allowed the researcher to recognize personal preconceptions of the research topic (Charmaz, 2006). The researcher compiled focused codes to determine categories and generate themes as they emerged from the data (Charmaz, 2006). The data was interpreted further as themes emerged, allowing the provisional categories to become a theoretical framework to describe the perceptions, experiences and understanding of the participants. Data collection and analysis continued until theoretical saturation was reached, and no new details describing a category emerged (Charmaz, 2006).

Data analysis.

The researcher began data analysis by listening to the interview audio and reading through the transcripts to check for accuracy. The researcher analyzed the transcripts line-by-line to identify open codes that consisted of participants' words and phrases. The researcher coded the transcripts line-by-line, which assisted in the development of concepts. As the researcher wrote memos throughout the coding process, personal reactions to the data, recurrent words, phrases and potential themes were identified. The researcher then typed the identified open codes into one Word Processing document, formatted with two columns. The compiled open codes resulted in a 60 page, typed Word Processing document of approximately 5,000 open and *in vivo* codes.

Throughout the data analysis the researcher utilized an iterative process to uncover the underlying implications, relying on memos to identify and construct meaning from the raw data to aid in the development of codes. The researcher maintained a running memo of repetitive words, phrases and potential themes. This memo document was condensed and labeled with 22 provisional categories. Twenty-two new Word Processing documents were created to separate and sort open codes, based on common meaning and repetitive words that emerged from the data. The researcher systematically moved open codes from the original 60 page Word Processing document to the associated focused codes document. The researcher changed the font to bold in the original Word Processing document to identify which open codes were sorted and moved to relevant documents. The researcher printed the 22 provisional category Word Processing documents and stapled them into packets.

The researcher subsequently reviewed the provisional categories to sort, separate and condense repetitive themes. An example of the collation of provisional categories originally included *importance of friendships, social media, siblings, social isolation, and self-care/coping*. The provisional category, *self-care/coping* was eliminated due to overlapping codes with the other categories, and instead, the researcher integrated the *self-care coping* codes into the relevant categories. These provisional categories were utilized in forming the theme *importance of connection*, supported by four defined focused codes *social isolation, siblings, value of friendships, and social media*.

The iterative process was beneficial throughout data analysis. The researcher sorted the data, developed open codes, condensed and compared open codes to define focused codes, and determined themes that emerged from the data. The findings resulted in six overarching themes each supported by four defined codes. The six themes that emerged from the data were: *CF Diagnosis, Degree of Familial Support, Impact of Life Expectancy, Prioritizing Health, Reality of Living with CF, and the Importance of Connection*. The researcher identified focused codes

relevant to the research questions which included: *infection prevention and control guidelines, social isolation, siblings, value of friendships, social media, and supported autonomy*. The two main concepts identified from the data that answered the initial research questions include *importance of connection* and *defining “normal.”* The researcher created a codebook to identify focused codes and their definitions, the themes that emerges from the data, and a sample quotation for each code (Appendix H).

Strategies for Ensuring Rigor

The researcher utilized peer debriefing and support along with memo writing to ensure rigor in this study; these strategies were also aimed to minimize researcher bias and reactivity (Padgett, 2008). The researcher wrote memos immediately following the completion of each interview to describe personal reactions, record observations and identify potential themes. The researcher reviewed the memos and documented decisions made during the analytic process to assist in identification of themes.

The researcher discussed memos with mentors and peers throughout the research study, including processing the interviews, data collection, coding and analysis to ensure accuracy and account for researcher bias. During debriefing, the researcher maintained confidentiality throughout discussions of the data, emerging codes and concepts. Peer debriefing allowed the researcher to obtain feedback which kept the researcher focused on the data during the analysis. Peer debriefing and support (Padgett, 2008), was obtained from both mentors, content experts, and peers, including:

- Numerous meetings, discussions and emails with Dr. Allison Werner-Lin, dissertation chair and an experienced qualitative researcher.
- Conversations with experts in the field of pediatric pulmonology, Dr. Michael S. Schwartz and Dr. Robert W. Miller, researchers and physicians who have provided care to countless patients with cystic fibrosis.
- Discussions with cystic fibrosis leaders and social workers at the North American Cystic Fibrosis Conference and Cystic Fibrosis Foundation sponsored events.
- Emails and exchanges with CF social workers throughout the course of the research.

- Conversations and emails with other grounded theory qualitative researchers from the University of Pennsylvania.

The above interactions were vital in supporting the reflexivity and transparency of the research, given the researcher's personal experience as a pediatric social worker. The ability to debrief with mentors, experts, and peers allowed the researcher to minimize personal bias while reflecting upon the concepts that were emerging from the data; similarities and discrepancies between participant's perceptions, experiences and understanding; the researcher's personal and clinical experience, and practice biases. These interactions minimized bias during the in-depth interviews with the participants, separating the researcher's professional experience working with children, adolescents, and young adults living with cystic fibrosis and other chronic illnesses.

Ethical Considerations

This study followed the ethical guidelines and procedures outlined by the University of Pennsylvania, including obtaining informed consent and maintaining confidentiality. The researcher received Institutional Review Board (IRB) approval prior to recruitment (Appendix A). The researcher successfully completed human subject protection training through the Collaborative Institutional Training Initiative (CITI Program) (Appendix I). Grounded theory methodology focused on using a systematic approach to analyze the data to generate concepts and construct theory (Charmaz, 2006). Following confidentiality requirements, the researcher removed identifiable information from the transcriptions. The findings are supported by quotations from participants to illustrate codes and concepts that emerged from the data (Charmaz & Bryant, 2011). To maintain anonymity of the participants, the quotations were described by their age or gender in the findings chapter.

Plans for Dissemination

Abstract Accepted – C. Toth (October 2016) “You wouldn’t know I have CF”: Cystic Fibrosis Social Connection and Isolation During Young Adulthood.” North American Cystic Fibrosis Conference, Orlando, FL.

Journal articles to be developed based on the above abstracts and specific findings, expected submission to a selection from the following:

- o Journal of Cystic Fibrosis
- o Pediatric Pulmonology
- o Family Systems and Health
- o Social Science and Medicine
- o Qualitative Health Research
- o Journal of Pulmonology and Respiratory Medicine

CHAPTER 4

Data Analysis

Organization of Findings

The purpose of this qualitative study was to explore the understanding, experiences and perceptions of social isolation amongst young adults living with cystic fibrosis. During the course of data analysis the focus shifted from social isolation to *social connection and isolation*. The findings of this study were derived from the analysis of semi-structured interviews with ten young adults living with CF. The findings are organized into two major sections, *Importance of Connection* and *Defining “Normal.”* These sections will include subheadings, direct quotations and phrases to support these concepts. Specific attention was taken to ensure verbatim communication from all ten participants to provide a breadth of perceptions, experiences and understanding.

Description of Research Participants

Sixteen individuals were initially recruited to participate. Four individuals never returned their consent forms. Two individuals submitted their consent forms, one scheduled an interview but later canceled and the other never scheduled an interview. No demographic information was obtained from these six individuals.

Findings are derived from interviews with 10 research participants, nine of the participants were female and one was male. Five participants were dating; three were married; and two were single. At the time of the interview, four participants had completed some college. Three obtained their bachelor's degrees: one of these three participants was enrolled in graduate school, and three earned graduate degrees. Three participants identified their employment status as full time, six participants were working part time: one also identified as a graduate student, and one participant self-identified as a college student. Six participants documented their

insurance as private; one indicated Medicaid as a secondary insurance, three participants had Medicaid as primary insurance: one participant indicated Medicare as their secondary insurance.

Participant's age at diagnosis varied from newborn to 15 years old. Five participants were diagnosed with cystic fibrosis when they were less than one year old. The other five participants were each diagnosed at different ages: three, seven, nine, twelve and fifteen years old. The participants called their disease "cystic fibrosis," "CF," "65 Roses" and a "genetic disease." According to the Cystic Fibrosis Foundation (2016), children have used the term "65 Roses" since 1965 to describe their disease because it was difficult to pronounce. The story originated when a four-year-old boy heard the name of his disease for the first time and pronounced cystic fibrosis as "65 Roses," (Cystic Fibrosis Foundation, 2016). It is important to maintain anonymity of the participants, and as such, in the findings presented below the participants will be described by their age or gender.

Importance of Connection

The theme *importance of connection* was emphasized and supported by the following findings. The *perception of social networks* described and explored how participants felt connected to CF and non-CF peers. Participants supported the discussion of social networks by interacting through *social media* to increase connection, though some participants chose not to connect via social media. Familiarity with the *infection prevention and control guidelines* impacted participant's ability to participate "face to face" with other individuals living with CF. These guidelines were explained to participants by their medical team, and several examples were related to hospitalizations and CF clinic appointments. These guidelines impacted participant's understanding of the *importance of connection*, and experiences of *social isolation*. Guidelines were also disregarded in the service of social connection.

Several participants described unique relationships with their *siblings* who were also diagnosed with CF compared to siblings not diagnosed with CF. All participants expressed the

value of friendships, and lessons learned regarding “real friends” and “sympathy friends.”

Finally, the findings described participant’s description of *supported autonomy* – relying on their family and significant others for continued support. The *importance of connection* was the key theme described in the findings related to the impact of isolation on young adult participants with CF.

Perception of social networks.

The participants explored the impact of their social networks in relationship to the recurrent theme the *importance of connection*. Several respondents felt adequately connected with social networks. A 24-year-old participant explained, “Having CF brought more friends in my life because I’m friends with a few people with... CF, too...it’s opened up that whole realm of connection.” She further described, “I’ve never felt alone or isolated.” The notion of connection was illustrated by many interviewees. Another 25-year-old participant mentioned inquisitively at first, “I don’t know that I’ve really ever felt isolated from other CF patients...” Upon reflection of her childhood she asserted, “...we were taught to be very independent.” Her family culture, level of maturity and self-assured independence reduced her considerations of isolation.

Many participants described their desire to connect, yet had minimal interaction with other individuals with CF. A 19-year-old participant explained, “I communicate more through people who deal with CF patients than I do patients.” As indicated, participants have a strong network communicating with their CF medical team. The aforementioned 25-year-old participant described:

While it would be great to talk to these other CF patients, at the same time it’s not a huge deal. I know I have CF and I know how I’m dealing with CF, so I don’t really need other CF patients in my life.

The notion of “independence” is elaborated on by the previous 19-year-old respondent, “I don’t really feel isolated at all... I don’t really need support.” Conversely, a 21-year-old respondent described self-imposed isolation:

I’m not opposed to participating... I really haven’t had time to get into it and respond... at another time, I think that I would become more active... if I had the time... I wouldn’t feel isolated as much. Right now, my isolation is my choice.

The participant expressed her self-awareness and disclosed “isolation by choice” from other individuals with CF as a way to indicate she had minimal need for the validation and normalization that come from connection to peers with similar disease-related challenges.

Participants described similar attributes related to their level of connection in their own terms.

Social media.

Participants shared their perception of feeling sufficiently connected to others with CF through the use of social media. Participants discussed being friends on social media with other individuals living with CF. *Social media* was incorporated by several participants that utilized Facebook and other platforms to spread awareness about CF beyond the CF community. They also provided and acquired advice, support, and connected to the CF community via social media. This allowed participants to actively or passively participate in social media at their convenience. However, some participants mentioned limited time to connect to other individuals with CF through social media due to other obligations including treatment, school, work and relationships.

The following participant reached out to others living with CF through social media as an outlet for support and connection, “I am in a Facebook group with all the people I go with to CF clinic and the same doctors as me, so that helps because we can still talk. It’s just through the internet.” Several respondents connected to other individuals with CF through Facebook, Instagram and other types of social media. A participant derived benefit from Facebook,

“because it’s the easiest and the most convenient” way to stay in contact. One 24-year-old participant described her reaction when she met another individual with CF, “I know what you’re going through, let’s be friends on Facebook.” She further explained, “I’m a member of a couple CF groups... mostly just looking for advice,” regarding treatment tips and high calorie recipes. Engagement, as a member of CF-specific Facebook groups allowed participants to deepen connection with others sharing disease burden. Another 24-year-old interviewee explained her frustrations when she felt overwhelmed, “if I wanted to really talk about it, I would probably go on the Facebook group.” She reported feeling disconnected from people without CF due to their lack of understanding pertaining to her life expectancy and treatment burden. She was striving for connection to others with CF who understood the day to day nuances and burden of living with CF.

Social media presented some participants with a double-edged sword; they could connect with other individuals who share their life experiences in ways that left them in control of intimacy so that they could control exposure to ‘worst-case scenario’ stories. A 21-year-old participant discussed, “I don’t really want to interact with people with CF right now, just because of my prior experiences... I was mostly just seeing things about death and dying.” She then mentioned, “I have never actually met anyone in person with CF... I don’t really want to... if I was going to reach out to someone with CF, it would be online or something, never in person.” The previous interactions on social media, as described by the aforementioned participant, led to her lack of “desire” to interact with other individuals living with CF because the participant was protecting herself from dialogue regarding life expectancy. Another 25-year-old interviewee chose not to connect to other individuals with CF, but she envisioned a more meaningful interaction in person than online. She discussed, “I’ll read blogs on the cystic fibrosis website all the time and I love hearing other people’s stories, but I don’t interact... at the end of the day I like to have a face to face conversation with someone.” She preferred reading about the experiences

of other individuals living with CF on social media, but was not comfortable posting her own story. She and several other participants would rather tell their story by engaging in a “face to face” conversation with another person, an exchange prohibited by the infection prevention and control guidelines.

Infection Prevention and Control Guidelines.

Respondents referred explicitly or implicitly to the *Infection Prevention and Control Guidelines*. The participants understood that the guidelines stated limitation that individuals with CF cannot be within six feet of each other. Some participants reported having an increased level of anxiety when attending CF clinic appointments due to the stringent infection requirements. Some respondents followed the guidelines warning to limit “face to face” interactions within and beyond the clinic, while others developed their own interpretation of the guidelines. The group that adhered to the guidelines respected medical staff for following the infection guidelines by gowning and gloving during interactions. These participants described experiences when they felt isolated during inpatient hospitalizations. In contrast, others personally disregarded aspects of the recommendations to follow their own rules.

Face to face.

Respondents remarked upon the impact of the *Infection Prevention and Control Guidelines* pervasive warning against “face to face” contact from other individuals with CF. They reported that their providers educated them on the risk of cross infection, instructing them to have minimal to no contact with other individuals living with CF. The implication of this for those living with CF is that they can acquire difficult to treat pathogens in their respiratory tract; yet limiting risk of cross infection left participants vulnerable to the social risk of limited contact. A 24-year-old stated, “It would be nice to talk face to face with somebody.” Another participant, age 24, stated:

It would be nice, though, to have a CF friend that you could hang out with because they would understand if you decided, hey I don't feel like doing anything today and hang out another day.

Another interviewee quipped, "The people who understand you, you can't spend time with." The guidelines challenge young adults' needs for intimacy in relationship with other individuals diagnosed with CF.

A 19-year-old respondent discussed his awareness of the guidelines while navigating relationships throughout school, and described a situation with a CF classmate. To make sure they did not cross paths at school events, they "got each other's numbers so we could tell each other where we were." The need to be aware of other individuals with CF for their own protection was described in association with the guidelines.

A few respondents mentioned seeking out or happening upon social exchanges with individuals with CF by choice. A 23-year-old participant described and defended a detailed "face to face" encounter with other individuals with CF while being hospitalized:

I met one of them... the one who was really sick... I did end up going and just saying hi to her, but I gowned up and put on a mask and stuff and kept my distance, but my doctor was not happy with us because of that... there was no contact, it wasn't something I was worried about... I felt like we were taking necessary precautions and it was ok. The three of us were all ok with it, so ultimately that's who it's up to.

She engaged in this interaction to reduce feelings of isolation. By reducing the physical barriers the participant created her own interpretation of the infection prevention guidelines, and balanced the risks associated with physical isolation and those associated with social isolation.

This participant further discussed the impact of the guidelines on her social development:

Growing up we never had friends who had CF. I know a few people but I don't ever see them. It's a texting relationship or Facebook, you just interact every so often, but not really a face-to-face relationship with anybody.

Even though these participants live in a generation connected by technology to social media, they still have a need for human connection. Some participants had friends with CF, but their interaction was limited to the use of technology. A female participant described her relationship with a friend who also has CF:

It's hard because we want to go out to eat or we want to go do something, but we can't be in a close, confined space, so we have to be outside or we have to just talk on the phone because we can't be in the same room for an extended period of time... it sucks because the people that understand me and know what I'm going through and that could be... my biggest support system... can't spend time with.

She went on to describe her desire for face-to-face communication with others, which included friends with CF:

I wish I could... be able to communicate face to face... I'm not a big texting person. I like talking to people. I like sitting down and having a conversation. It's hard that we're not allowed to do that, not that we're not allowed but there's big precautions and big consequences that could come out of it.

She understood the possible consequences of face to face communication with another individual living with CF. Repeatedly, she expressed her concerns on the impact these limitations had on her relationships with CF friends, and reiterated the need to connect beyond technology. Ultimately, she yearned for physical connection to others with CF.

The following 20-year-old participant described her experience feeling isolated from a cousin with CF due to parents following the infection guidelines. She described feeling somewhat connected to her cousin through Facebook and elaborated on the challenges of family gatherings, "going to the family's house during the holidays... could be around everybody else but not him... we're a little bit isolated from each other." She reflected upon multiple incidents and described feeling depressed after family gatherings. Human connection is important in all aspects of life, but within families this need to connect "face to face" is magnified.

Hospitalizations.

Hospitalizations are common for children, adolescents, and young adults with CF due to disease complications or scheduled “tune up” admissions to maintain their current health. Time spent in hospitals further separated participants from non-CF peers. Cystic fibrosis has rigorous treatment protocol, yet these implications varied among participants with particular respect to hospitalizations. Some participants described strategies to avoid the hospital, while others acknowledge the necessity of hospitalizations.

Participants discussed their understanding of the differences between types of hospitalizations. Scheduled “tune up” hospital admissions are performed to improve and maintain their current health. Unscheduled hospital admissions were related to respiratory exacerbations, hemoptysis, or significantly decreased lung function. Participants alternatively described the negative implications that coincided with unscheduled hospital admissions. A female participant described her perception of her health declining correlating directly to the doctors directly admitting her to the hospital, “when I was younger I never planned them because I didn’t want to... when I got sick enough they [the doctor] would throw me in the hospital.” Several participants discussed the frequency of hospitalizations throughout their lifespan and the intensive treatment regimen they experienced during their hospital admission, which included airway clearance therapy at least four times a day.

Participants discussed the negative implications of their hospitalizations, during which they felt more physically and socially isolated due to stringent infection precautions. CF participants were required to stay in their hospital room and to wear a mask when leaving the room. An 18-year-old participant described feeling isolated in the hospital:

I would go into a hospital... and I couldn’t leave my room. I couldn’t go anywhere near the door... I felt pretty isolated...I already felt isolated enough... I could never really leave my room.

While the participant was allowed to leave the hospital room, it was only under the condition that they wore a mask and were accompanied by a parent or medical staff personnel. Individuals with CF are required to stay in their hospital room during the admission unless being transported to diagnostic testing. To alleviate some symptoms, the respondent would follow infection protocol so that she could leave her room:

I would gown up, put a mask on, put gloves on, put everything on I needed to, and I would try to walk the halls a little bit if it could help me feel not so isolated.

To reduce feelings of isolation a 23-year-old participant shared advice to others with CF, “making sure that you do everything you can do to stay out of the hospital.” She reflected upon being hospitalized during her childhood and receiving letters from classmates:

It was really nice, but at the same time, it had this strange effect of making me feel more isolated because I could just tell from a lot of their letter that most of the kids either didn't know why they were writing to me and didn't know what it was I had or didn't care.

This thoughtful interaction from her elementary school classmates had an adverse effect on this participant's wellbeing, highlighting the divide between herself and the world outside of CF. When hospitalized, the participants' world revolved around medical staff, family members, significant others and occasionally friends.

CF clinic appointments.

Many participants were aware of the *Infection Prevention and Control Guidelines* and discussed wearing a mask at CF clinic appointments, in particular. Several participants described the impact of these guidelines in contributing to feelings of stigma and isolation. As previously mentioned, several interviewees respect their CF medical team by following the guidelines. A few participants remarked on their experience during CF clinic appointments when their medical team wore gowns and gloves. Knowledge of the guidelines and risk of cross infection was a

source of anxiety for some participants, especially at CF clinic appointments. A few participants recalled upon their experience attending a CF clinic appointment, and reported feeling segregated when they arrived and immediately had to enter the exam room. A 24-year-old interviewee mentioned, “When you go to the doctors and you have to go right back to the room with your mask on and you can’t come down the hallways if somebody else is out there.” The participant shared her experience when “somebody else” with CF was in the hallway at the clinic she had to remain in the exam room. The immediate attention provided to this population by medical staff at CF clinic appointments increased feeling disconnection from others, making an invisible illness suddenly visible.

Several participants, however, did feel comfortable when doctors and medical staff gown and glove when entering the exam room. A 23-year-old participant described her knowledge about the guidelines, “people with CF aren’t supposed to be within close proximity of each other for risk of passing lung infection or bacteria back and forth... be really cautious when you’re around other people with CF and avoid it if possible.” This participant had a clear understanding of the guidelines and the ramifications from interacting with another individual with CF. Another 21-year-old participant discussed feeling nervous when going to CF doctor’s appointments:

My doctors always gowned down when they come into my room. The last doctor I was at is a new doctor, and when I went to give him a handshake, and he instead did an elbow bump... I always get a little nervous when I go to the doctor, because I know a lot of CF patients here... I only get nervous because the infection control thing.

The participant described feeling nervous when attending a CF clinic appointment due to the risk of cross infection from other individuals with CF.

Sibling relationships.

Participants described the importance of the relationships with their siblings. Some participants had siblings also diagnosed with CF, while others had siblings not diagnosed with CF. The difference reported by participants with CF siblings was that growing up, the CF

siblings understood the disease and required treatments. In contrast, several respondents with CF siblings perceived significant differences during the transition of care from parents to individual responsibilities, including acceptance and disease management techniques. Several participants described the relationship they had with siblings not diagnosed with CF, in which the non-CF sibling relationship was encoded with perceived jealousy and misunderstanding. Participants expressed varied experiences related to their level of connection to their siblings.

CF siblings.

Family members with CF are a unique population, as exhibited by several respondents. Some participants had siblings also diagnosed with CF and understood the infection guidelines recommendation to separate individuals with CF 6 by feet, did not apply to them. A 21-year-old participant discussed feeling nervous around other individuals with CF aside from her brother, “I know I would never interact with other CF people, physically within real life, except my brother has CF.”

These respondents have developed a unique perspective, but the relationship between CF siblings also had distinctive challenges. A 20-year-old interviewee explained that her CF sibling went through a rebellious phase. This “negatively” impacted their relationship due to conflicting disease management techniques and inherently increased feeling disconnected from each other.

She further elaborated:

She is in that rebellious phase that I was in, and I just want to shake her... do your treatments, do it right, do your treatments, take your pills, let's go exercise and get our lungs up... I don't want her to go through that phase as long as I did because I know, in the long run, it negatively affects me.

The participant described learning from her rebellious phase and understood the negative health implications from not following her prescribed treatment regimen. She yearned to share her experience with her sibling also living with CF, however, her sister was not receptive to the

information. Older siblings with CF discussed life lessons on their journey of disease management and described the protective nature within their CF sibling relationship.

A 23-year-old participant with CF described her unique situation having two other relatively healthy siblings with CF, “my brother and sister also have CF. ... The three of us grew up together all in the same household all having CF.” She discussed the decline in her sister’s health which caused increased stress on their relationship:

Luckily for her, she’s been relatively healthy her whole life, it’s just started to impact her lately... it’s just added stress because she’s so defiant and refuses to take care of herself, which is sad... adds stress and worry... hopefully she turns around

The CF sibling relationship is complicated; the lack of disease acceptance and non-compliance to prescribed treatment could lead to worsening disease severity and possibly death. These negative health outcomes of a CF sibling could potentially lead to the other CF sibling envisioning their own future. A 21-year-old participant described the strained relationship with her CF sibling and their different styles of disease management. She shared feeling disconnected from her CF sibling and the choices he made regarding his health:

He rebels against his treatments, he doesn’t like to do them... he wishes he didn’t have to do them... I think it’s really hard for him to accept that he has CF... it’s really hard for me to watch him get sick and not be able to do anything about it.

As discussed by participants, the once robust relationship between siblings diagnosed with CF can be hindered by differences in disease acceptance and disease management techniques.

Non-CF siblings.

Young adults living with CF discussed having strained relationships with their siblings not diagnosed with CF. A 24-year-old interviewee felt remorse and disconnected from her non-CF sibling. She discussed self-imposed blame for the negative attention-seeking behavior of her non-CF sibling. This participant felt increasingly sequestered due to sibling jealousy and

rebellion, “I think she feels as though she is not as important as I am, to my mom and dad, and I don’t want her to feel that way.” To alleviate these feelings, the participant mentioned she would “spend time with her so that she knows I love her.” Participants with non-CF siblings described how they spent time together to try to improve and repair their strained relationship. An 18-year-old interviewee described the strained relationship with her non-CF sibling due to her parents’ involvement with her having CF:

When I would get really sick and my parents would have to pay more attention to me, he would feel left out and he would get oppressive and he wouldn’t be happy. It really affected him... I think it actually really impacted him... more attention was always paid to me because of the medications.

The participant’s description of her family resonated with other respondent’s description of non-CF sibling’s negative attention seeking behavior, due to lack of understanding and the perception that their parents paid more attention to their CF sibling. The perceived negative emotional impact on the non-CF sibling did not go unrecognized by the participants. The disease impacted the respondent’s family system and non-CF sibling relationships due to time spent with parents, treatment burden, frequent illnesses, and recurrent hospitalizations.

Another 25-year-old participant described an incident that occurred with her non-CF sibling. The desperation from the non-CF sibling led to the participant to acknowledge her own loneliness and symptoms of depression. The respondent reached out to her non-CF sibling and had an “honest” conversation, “I honestly never went to anyone and I really actually didn’t even start to really address it until... my brother, actually started that way himself.” This exchange deepened their relationship and alleviated symptoms of depression for both siblings.

A 25-year-old participant described feeling guilty living with CF and the negative impact it had on her family. However, she described inspiring her younger sibling:

She wants to be a cystic fibrosis doctor, and since she was 7 years old, she has told me that she's going to cure me. I do feel really good about that side of my family relationships, because I inspired my baby sister, and that's so meaningful to me.

The participant balanced the negative implications of living with CF with the positive effect it had on her younger sister. Participants acknowledged the burden of CF on non-CF siblings, which included the non-CF sibling often being overlooked due to extensive treatment regimens.

The respondent further elaborated on a scenario where her non-CF sibling felt guilty:

She figured out that what's happening is that her sister is actually dying and her sister is actually struggling, and she feels guilty because it wasn't her... made me so sad because I never realized that my little sister felt guilty that I was the one with CF and not her. She wishes that it was her so that she wouldn't have to see me struggle, and I'm very happy it's the other way around and I don't have to watch her struggle.

The interviewee explained being "happy" or relieved that her siblings were not diagnosed with CF, because she understood the daily impact, "struggle," and negative implications the disease had on her family.

Social isolation.

CF had a significant impact on participants' social life. Several participants felt isolated from individuals in general, due to their lack of compassion and understanding. *Social isolation* was defined by participants through utterances of frustrations that they were not understood and felt disconnected from others. Participants weighed the impact of isolation from others in general and lacked feeling isolated from others diagnosed with CF specifically.

Participants were self-aware regarding the impact their disease had on establishing connections. A 25-year-old respondent affirmed that she did not feel separated specifically from the CF community – the seclusion was from "individuals in general." Several participants considered the impact of isolation from non-CF individuals and felt indifferent regarding the connection to others with CF. A 23-year-old participant noted, "there are definitely people who

just get it, and some who don't." The findings demonstrated the participants desire to maintain their health and routine to increase connection with others.

Several participants felt increasingly disconnected after disclosing their diagnosis to others. Participants reported recurrent feelings of isolation from individuals in general, especially non-CF peers. A 25-year-old participant expressed, "I have felt isolated from the general public a lot." Another 25-year-old participant mentioned feeling secluded from the general public due to lack of awareness regarding cystic fibrosis, "I think it's one of those illnesses that just gets pushed off to the side, because not many people have it, but it's still out there." The iteration from participants after describing CF was similar, as a 24-year-old participant described:

I always like to mention, too, that I don't want you to feel bad for me. Because usually after you tell someone this, you see the look of pity in their eyes of, oh my God, I just made her talk about this, and oh my gosh she has to deal with this. And it's one of those things, it's like, no, don't – I don't need pity for it. I'm happy and I'm healthy and I'm doing what I need to do and what I want to do.

Participants described living with an invisible illness, and when this participant informed a stranger of her diagnosis the individual displayed looks that exhibited "pity." This participant, along with several others, acknowledged the need to continuously defend their health.

Participants expressed the recurrent notion to inform the public that they were "healthy," despite disclosing their disease or coughing in public. Participants discussed constantly having to inform family members, non-CF peers, their medical team and individuals in general others of their health status.

Participants felt isolated from others in general due to their lack of understanding. A 23-year-old participant explained, "not to assume that everybody who has CF is going through the exact same thing because it can be very different." Respondents expressed that they look like everyone else and do not look sick. Participants' experience of social connection was linked to acceptance by their college peers, and as one 19-year-old participant generalized, "most CF

patients don't like to be looked at as having an issue." Coughing in public continued to be a struggle for participants where they felt judged by general public. A 25-year-old participant elaborated:

Not having to worry all the time about catching something or having a coughing fit in public and getting weird looks from people. I just want to fit in. I don't want to stand out because of CF... it brings negative attention to me.

She further described a situation she encountered, in which she was unable to inform the individual about her lung disease:

I was coughing in public and somebody gave me a dirty look... Don't judge me. I have this lung disease, by then they're gone and you're getting stuck with this feeling of people are just judging you constantly.

Coughing was a symptom described by several participants that brought unwarranted stigma from individuals in general. Another 24-year-old female discussed, "I would want people to know is that we cough but it is not contagious and we're not always sick because we are coughing." Participants described wanting to be accepted as a human being regardless of their physical appearance or recurrent coughing.

Bullying.

An 18-year-old participant described an incident in middle school during health class where she discussed CF as a genetic disorder. Her middle school health teacher stated in front of her classmates, "don't people with that disease die young?" This comment about her life expectancy increased bullying by non-CF peers, and caused her to feel disconnected and afraid to go to school.

One interviewee briefly mentioned being bullied due to her body shape and small build; she experienced insults such as "chicken legs," compared to her barrel chest and abnormal shaped abdomen. Another 25-year-old participant discussed a series of bullying incidents that

occurred during high school. One incident occurred when she was running the track in gym class and non-CF peers made fun of her breathing. She described, “I was just breathing how I breathe...that’s the first time I encountered bullying... there are just so many times I can think about.” She further discussed bullying related to an instance where she felt segregated for having CF. She mentioned having an extended illness where she had to leave the high school soccer team:

I actually had to leave the soccer team because the girls on the soccer team decided that I was just faking to get out of having to play... I felt like I was not really being accepted with my issue.

Another 24-year-old participant felt bullied and disconnected from her friends when they didn’t understand the impact the disease had on her daily life:

My friends when they would do stuff and I don’t want to because I’m tired, or I don’t want to because I want so sleep... my friends know that I have CF but I don’t think they know, you know really know.

Key points mentioned by participants were regarding feeling accepted and understood by non-CF peers. Participants felt their connection to others increased when others were sensitive and aware of their disease.

Smoking and social seclusion.

Feeling isolated in social situations was a factor mentioned by participants. Respondents discussed the impact of smoking as an assault on the lungs for individuals living with CF. Participants described the prevalence of seclusion in areas where individuals not living with CF might barely notice. No participants reported smoking themselves, however, several participants elaborated on the impact of smoke from their friends and the general public. A female participant’s family members were even unaware or disregard the warning of smoking around individuals living with CF. She further elaborated, “I have a family that smokes... secondhand

smoke is just as bad.” Smoking induced respondents’ stress levels and increased feelings of isolation. A 25-year-old respondent discussed hiding CF during college:

I really hid everything, I didn’t always feel like I was getting the college experience a hundred percent especially interacting with people because I didn’t want them to kind of be like, oh well, she’s got to do this instead of hang out with us as a group or something like that... because I had to think about my health first. A lot of college parties you are smoking... I just didn’t get to experience that part of it.

The aforementioned participant explained that she felt segregated from non-CF college peers due to the prevalence of smoking at college parties. Another 18-year-old interviewee also commented on the challenges experienced during freshman year of the college, “I don’t drink, I don’t smoke, I don’t do any of that... for the first few weeks I was by myself.”

A 24-year-old participant described the impact of smoking, “if we go somewhere and somebody’s smoking, it’s twice as worse for me than someone else just because of the smoke.” A 20-year-old explained feeling detached from her friends when they were recurrently trying to smoke around her. She stated she was “continuously reminding” her friends, “you can’t smoke around me, you can’t smoke in my car” The participant expressed her frustrations with her friends’ lack of awareness and insensitivity to her wellbeing. Another 25-year-old female participant described a scenario where her friends invited her to a bar and she initially felt included:

It’s had a social impact on me that I’ve noticed I’ve had to be very careful about which friends I pick and where we go... we’d get to the bar and they’d sit on the patio and start smoking, and I’d have to leave.

Being included in activities allowed participants to feel accepted by their friends. However, participants felt increasingly isolated when their friends disregarded the negative health implications from smoking around them.

The aforementioned participant expressed that she was adamantly opposed to people smoking in public, because it has drastically increased her feeling segregated on multiple occasions. She described a situation where she left parades due to individuals smoking outside:

I've actually had to leave parades before because people think... it's fine to smoke. It's not going to bother anybody, but it really does and it would get so bad.... I had to leave for the smoke. That's been very frustrating, trying to tailor my social life and where I go to if there's gonna be smokers or not... it never occurred to them that smoking outside could be harmful to somebody... that made me feel very different... people don't take a moment to consider what I'm going through.

Several participants felt different and misunderstood due to negative health repercussions of other people smoking in public.

Value of friendships.

Participants described the *value of friendships* related to two different types of friends “real friends” versus “sympathy friends.” The difference between good friends and acquaintances was determined by “who’s worth keeping around.” Several respondents described the importance of having friends that understand the intricacies of living with CF. Some participants were bullied by non-CF peers. Other participants described the impact their treatment burden had on their social lives.

Making hard choices: Socializing with friends or completing treatments.

Time consuming treatment regimens impacted participants’ friendship, and on multiple occasions, participants reflected upon the effect CF had on their childhood friendships. They discussed lack of participation in age appropriate activities such as sleepovers and playing sports, due to breathing treatments. One participant noticed the impact of her treatments during high school. She described in, “high school I really started to notice it... I’d be at my friend’s house and my mom would call.” The participant elaborated on the conversation she had with her mom regarding a sleepover, “I’d have to come home and I’d have to do my treatments, I’d have to get my meds... I just wanted to go. I wanted to have fun.” The notion of having “fun” versus

completing time consuming treatments was discussed by several participants. Another 20 year old female interviewee expressed her frustrations:

I just remember growing up, it was always frustrating because I couldn't go to sleepovers at certain points because I was either starting to get sick or started showing signs of my lungs getting too congested, so I would have to do extra treatments.

Several participants felt frustrated and left out of age appropriate peer activities due to their treatment burden and declining health.

Female participants discussed the impact of parental influence on their health maintenance. Parents reminded participants to complete time consuming treatment regimens, which led respondents to spend less time with friends. Conversely, some participants skipped treatments to spend more time with friends. One 25-year-old participant reflected upon an entire year when she did not complete any treatments to reduce feeling secluded:

I was sick and tired of doing my treatments...I didn't want to do my vest. I probably actually didn't do it for a straight year. I really wanted no part in even taking a simple antibiotic orally... now I am dedicated to the point where, if I skip a vest treatment, the next morning I am doing double.

She elaborated that she now understands the importance of balancing her treatments to reduce these feelings of seclusion and decrease the risk of hospitalizations. A 24-year-old skipped treatments as a teenager, "I didn't want to waste my time. I didn't want to come home early or wake up early before high school doing them." During adolescence this behavior allowed her to feel more connected to her friends. The participant further discussed being worried about the future, about health and about getting sick:

Finding the time to get all my treatments in and if I feel like I don't have enough time then I worry that I am going to get sick and my biggest stressor probably is that I am just going to have to drop everything to go into the hospital just to get better when I would much rather plan it.

It was important to many respondents to balance health, disease management, and relationships. “Finding the time” to complete treatments was essential to managing their health, however, socializing with their friends was a personal priority.

“Real friends.”

Participants understood the *value of friendships*; several described their friendship journey. A female participant expressed gratitude, “I’m very lucky to have found a group of friends that are supportive and that are understanding of my condition.” She reflected upon a time when she learned the importance of friendships,

There have been a few times we’ve had issues and I’ve actually had to cut friends out of my life because of how they were.... I actually stopped being friends with him because of how isolated he made me feel.

She described a situation when she “felt special” that her “real friends” supported her despite adversity. She discussed her, “real friends went out of their way to make sure that I was included.” One 18 year old participant highlighted the lesson of quality of friendships taking precedence over quantity of friendships:

Not able to spend a lot of time with friends because between the homework, work, school and all my treatments, I don’t really get any time to go out and actually spend time with friends, so it’s been really hard to make friends my life. My entire life I have managed, but most of them have to understand about the CF and all the treatments. I have one really good friend.

The participant described the difficulties developing and maintaining friendships due to balancing her health with other responsibilities. Respondents that discussed having at least one good friend helped them feel more connected to others.

Another female participant described having understanding friends, “they’ll be there for you if they’re good friends, and if they don’t stick with you then they aren’t good friends.” The

quality of friendships allowed participants to deepen their connection and reduce feelings of isolation. A 23-year-old participant described her friendship journey:

I've always kept friends who understand it and care about it... I don't waste my time with people who don't understand it or don't care or are kind of intolerant to it. I have a good set of friends who are understanding... it's a learning process throughout my whole life and just learning who genuinely cares and who's worth keeping around.

A 20-year-old female interviewee described the difference between friends and acquaintances, "my best friends, they'll be there, they'll text everyday if I'm sick... how are you feeling today?... versus... They keep on saying I'm sorry. They keep asking how it happened, what's wrong?"

Young adult participants described CF to their acquaintances, but found they lack the understanding that good friends have developed. A female participant described:

Friendships, the most difficult part for me...is being open about it...my best friend is supportive, but when it comes to new friendships I am a lot of times very cautious to talk about it.

This participant described how difficult it was to disclose her disease when meeting new people, which hindered developing new friendships. Conversely, a male respondent openly explained CF to his friends. He described it as, "A better way to open the conversation to get to know people better... I guess I'm more proud to have it...I would definitely not be the same person I would be – I am today if it wasn't for CF." This participant accepted CF and developed a sense of personal pride. CF became integrated as part of the participants' identity, as they learned through trials and tribulations how to disclose their disease and develop "real friends."

"Sympathy friends."

Respondents learned the importance of healthy friendships versus "sympathy friends." These "sympathy friends" were initially thought to be "real friends," but throughout their relationship, participants learned that this type of friendship was established because they had cystic fibrosis. Many participants found good friends and recognized the importance of

maintaining these relationships, while others described scenarios where their “sympathy friends” were only their friends due to their disease. An 18-year-old participant described her history with “sympathy friends” and how she has found “real friends.” She discussed:

I don't want sympathy friends. I don't like when people feel sorry for me, which has happened before. I've gotten sympathy friends where they use me and they tell other people... my best friend's dying... I've figured out how to weed those out and get friends that actually care about me, not just my story... CF doesn't make me. I want people to know me before they know that, because I'm not CF.

This respondent wanted to be known for who she was as an individual and not just for her disease. Another female participant strongly affirmed, “I don't let CF define me.” CF was incorporated into the respondent's identity, however CF did not define her.

Many respondents reported that they have been bullied on several accounts. An 18 year old female participant described being bullied for several years in school, but kept her “sympathy friends” for fear of loneliness:

I was picked on, I was kind of bullied... I didn't have a lot of friends, and the friends that I did have were sympathy friends. But I didn't really want to weed them out because I didn't have anybody else.

To combat these feelings of disconnection from their “sympathy friends”, participants described relying on their family members and significant others for support.

Supported autonomy.

Several participants described *supported autonomy* throughout their interviews, characterized by developing independence while they remained attached to their family and significant others. Participants identified being supported by their parents and significant others. Some participants wanted decision-making capacity yet hesitated to ask for help. An 18-year-old participant described her family as her “biggest support system, if something is wrong they are there. If I don't feel good they are there to make sure... I get to all my doctor's appointments.”

Respondents described the support from their parents or significant others while attending medical appointments and visiting during hospitalizations.

Supported autonomy allowed participants to develop connection to bridge the gap between family and friends. A 24-year-old participant discussed feeling supported when her mother or husband accompanied her to CF clinic appointments to provide support. She described traveling together in the car for several hours to attend her CF clinic appointments, “even to this day my mom will go with me. She’ll take off work and go... if my husband can’t go.” She described the *importance of connection* from supportive family members and her significant other. Another 25-year-old participant’s perspective shifted when she met her husband. She expressed, “it wasn’t until I met my husband that I actually started taking my medication... I can handle anything that happens.” She mentioned the importance of her husband making her feel less isolated, more conscious about her health, and held accountable.

Participants also described the importance of having an emotionally supportive family. An 18-year-old stated, “now that I’m living on my own and having to take care of myself, it’s a little harder.” She described this transition to independence while being supported by her family. A 23-year-old participant embodied the notion of *supported autonomy* as she described her transition to college and becoming “a real life adult;” during which, she still relied on her mother for support with her CF care, “wanted to do that on my own and still kind of wanted her help.” She discussed being married to a supportive husband, “from day one has gone to all my appointments with me and just gets it... understands it and knows how I want to live my life with it.” Another female participant echoed the previous respondent’s remarks related to her supportive significant other, “he is always there for me if I am sick and when I am not sick... very helpful to me, to have someone to talk to and lean on.” Having supportive significant others was important to participants’ physical and emotional wellbeing.

One of the challenges noted by participants was the knowledge of life expectancy for the CF population. Participants relied on their family and significant other for support to cope with the looming life expectancy statistics. One participant described, “The only thing we can do is live for the moment and life for us.” Another female respondent elaborated, “That number’s a lot closer and sooner than it is for a lot of people... get overwhelmed with it.” The relationship explored by this participant segued into relying on her husband for support to discuss anxiety related to disease progression and life expectancy. A 25-year-old described leaning on her family and significant other to minimize symptoms of depression and life-limiting knowledge of the constant reminder of her life expectancy, “I spent my whole life thinking I was gonna die, and here I am and I’m not dead.”

Defining “Normal”: “I don’t want to stand out because of CF”

All participants, without probing, expressed how they work to feel normal in their lives, making this a powerful iterative finding. The researcher identified the implicit need for the participants to discuss further, and thus explored participants’ meanings of normal. To arrive at their own, unique, definitions of “normal,” participants compared themselves to non-CF peers related to working, going to school, relationships and health concerns. A 20-year-old female interviewee discussed: “A normal person doesn’t have a lot of worries when it comes to health.” Another participant related “normal” to developmental milestone in comparison with non-CF young adults that were going out with friends and moving away from home. One 25-year-old young woman eloquently expressed, “Everyone has something that’s not normal about their lives.”

Young adult participants with CF described the yearning to be able to fit in and adapt to their environment. The interviewees discussed instances when strangers acknowledged their differences related to coughing in public or commented on their physical appearance. This acknowledgement and perceived judgment caused the participants to feel stigmatized in society.

A 19-year-old male participant discussed, “Most CF patients don’t like to be looked at as having an issue.” Female participants in their twenties elaborated on this notion: “normal is just you can have attention, but I don’t want the negative attention from my health” and “fitting in, not being singled out. You’re comfortable in your own skin. Like to stand out for being outspoken or being really smart... don’t want to stand out for being sick.”

These participants described what life is like living with an invisible illness, “You wouldn’t know I have CF just by interacting with me or looking at me.” Participants discussed their appearance compared to non-CF peers, “no one would know that I have CF.” Another female participant explained, “You look at me and you have no idea that I have anything wrong with me.” These participants experienced stigma when others in the general public noticed and commented on the differences. A 25-year-old female participant describes her rebellion against the guideline related to wearing a mask at her clinic appointments due to her perceived normalcy:

Because one good thing about cystic fibrosis is that if I really want to, I can blend in with everybody else. Look at me in a line-up and you will not know that I am the person who’s sick. You won’t know. I can look so normal... I always refuse to wear the mask because I just like to feel normal. I just want to be normal. I’m trying so hard... When you’re in the clinic, it’s like maybe I’m the sick person, maybe I’m waiting for the sick person. Nobody knows. And that makes me feel like I’m a little closer to normal. Like maybe I’ll just blend into the background.

The participant described the perceived impact from wearing a mask. A 21-year-old respondent echoed this notion, “I don’t wear a mask... I just don’t want the looks... I want to fit in and feel as normal as possible.” If these participants wore a mask they would not feel “normal” because the mask related to sick stigma in society. Several participants, like the respondent above, discussed rebelling against the *Infection Prevention and Control Guidelines* established to protect individuals living with CF from the risk of cross infection. Yet, these markers identify people with CF as sick.

Social comparison as normal.

A common theme within the reality of living with CF was the challenge of living a “normal” life. The definition of “normal,” as described by the participants, was personal and unique, and included variants within the definition. Some participants viewed “normal” as a form of social comparison to their non-CF peers. Participants articulated that “normal” is marked by the absence of medication, fewer hospitalizations and less health concerns than non-CF peers. A variation of “normal” for participants can be defined by the absences of CF related symptoms. One 25-year-old female discussed “normal” related to her health status, “I know I’m not sick when I can laugh without coughing.” She saw “normal” as a constant flux that depended on the severity of her symptoms and her capacity to react naturally to her surroundings without physical consequence for her health status. For some young adult participants “normal” is a process defined in contrast to non-CF peers. For others it’s the capacity to seem normal to an observer.

Many participants expressed feeling “normal” living with CF, “I actually think that my life is pretty normal.” Another participant described, “I’ve been able to live my life as normal as possible,” seeing “normal” as a goal to achieve by minimal observable health events and the least amount of complications of disease intrusions. Compared to other peers, this participant mentioned, “I just live my life what I consider normally.” The following participant lived his life feeling that CF wasn’t a game changer, “I really try not to make it any different than living without it.” For this participant, decisions weighed social and developmental choices over health demands whenever possible. Some participants chose the appearance of living “normal” as a priority over maintaining their health.

Individuals living with CF have extensive daily treatment regimens. The young adults interviewed with CF focused on the importance of maintaining health and their daily routines. Several participants described the impact of treatments and that on several days, normal consisted of “sheer functioning,” characterized by the ability to get out of bed and go to work or

school. Participants related the definition of normal to health and medical treatments, “My definition of normal is not having to get up and do treatments and all the medicine.” Another female participant expressed her definition compared to non-CF peers, “Someone who is in good health. Someone who doesn’t have to do breathing treatments or take vitamins in the morning and night and doesn’t have to do breathing treatments, have tune-ups, have PICC lines and have IVs.” Participants further discussed steps taken to maintain their health, which included hospitalizations requiring a peripherally inserted central catheter (PICC) line and intravenous antibiotics.

As previously discussed, young adults with CF understand their disease and the possibility of getting sick. The knowledge of such – disease progression and life expectancy – was a worry shared by the young adults interviewed. A 25-year-old participant related being “normal” to not having to worry, and mentioned, “Not having to worry all the time about catching something or having a coughing fit in public, and I just want to fit in. I don’t want to stand out because of CF.”

Anxiety and depression symptoms.

Several participants discussed their symptoms of anxiety and depression. A female participant discussed the mind-body connection, stating, “If you don’t physically feel well that can impact your mental health drastically.” She understood the impact of mental health on her disease management, treatment adherence, and overall physical health outcomes. Several participants described a history of having suicidal ideations. A female interviewee described a time when, “I was okay where if I didn’t wake up the next morning, I was fine with that.” This respondent recognized “that was definitely my darkest point ever,” and further explained she was “never screened for it [depression].”

Another participant supported this notion of mental and physical health, “they’re definitely related.” She also mentioned the “stigma that comes with saying... you have

depression.” A 24-year-old participant elaborated on worrying, “I feel like I don’t have enough time when I worry I am going to get sick.” The extent of her worrying went beyond CF. She mentioned her treatment regimens infiltrated school, work, and time spent with her family, friends and significant other. Young adult respondents explored topics surrounding anxiety, worry, depression and suicidal ideation. A female participant described, “The things that I have to worry about...no one else does.” She expressed that she worried about disease progression and her CF getting worse, meanwhile, she perceived that her family and non-CF peers did not have the same health concerns.

Screening for anxiety and depression.

Young adult participants were not forthcoming when they described mental health symptoms to their CF medical teams, either due to shame, fear of stigma, or because they didn’t want to disappoint providers they felt close with. A female interviewee commented on her CF medical team screening for depression, “they always kind of ask you, just as a passing question.” She further discussed her reluctance to answer:

It’s always awkward because my mom was either there with me or my husband... I think I’ve felt it [depression] before, but whenever they ask, I just say no, because I don’t want to come off as self-pitying or that it’s something that you dwell on... and it’s not a comfortable question to answer for people, to answer honestly.

She described her self-awareness and later stated, “It’s something I would imagine probably goes... under diagnosed a lot in people with CF. One, because they don’t recognize it maybe, or two because they’re not comfortable saying that they feel that way.” Another female respondent expressed concerns when asked about depression by her medical team; she stated they were not “as careful about it as they should be.” She worried that her medical team would enter the exam room and ask, “I see you put on here that you’re feeling depressed, what’s that about? And then I would be... caught.” She later stated, “If I were to do something about it [depression] I would go elsewhere.” Participants felt disclosing their anxiety and depression symptoms would make them

feel stigmatized by their CF care team. A 25-year-old participant described a situation when she did not disclose her symptoms, even when asked by her CF medical team:

I was never open about it either, but I was at the point where I felt kind of ashamed to even say something, because I know that I would never be able to actually go through with it, but even me saying that, I know now that yeah that was pretty serious.

A 23-year-old interviewee explained, “You see your CF team so often, and you don’t want them to see you or think of you that way... I would go to a psychiatrist or something.” Her experience echoed other respondents that felt too close to their CF medical team. Another female participant reflected upon her symptoms of depression and anxiety, “look back in disbelief mental health got that bad.” After years of being bullied in middle school, the aforementioned participant reflected upon her “severe depression” during high school:

I didn’t really fit in and my freshman year of high school... I went into severe depression... I had no friends and the friends I did have – I was having suicidal thoughts – she told me she didn’t want to be my babysitter... so that really stuck with me.

This participant had suicidal thoughts and felt abandoned by her friends. Her family recognized the severity of the situation and the participant went to counseling. The participant described her counseling experience, “I saw a counselor... during high school and she didn’t really help a lot. She just told me that all of my problems are centered around the fact that I have a chronic disease.” She described advocating for herself when she informed the counselor that her depression was not “centered around” her disease, “I’m just having trouble finding myself and I need somebody to listen, for me to talk to, but she kept turning it around and making it about CF.” She explained that her counselor did not understand the complexities of her disease and when she went to counseling it made her “thoughts of CF worse.” This participant later described how she recognized, “I just needed somebody to listen, not to judge me and not have a biased opinion.” Participants described withholding their symptoms from their family, friends, and health care providers to appear “normal.”

Balance and the pursuit of normal.

The young adult participants persevered despite their disease. The participants have obtained advanced educational degrees, as well as part time or full time employment. The inability to balance employment and their disease is a reality for some young adults with CF, as noted by one 20-year-old participant, “I can’t really work the way other people do.” Conversely, a 24-year-old female participant working full time described her “normal” compared to individuals not living with CF: “My normal is getting up, going to work and then doing my treatments and all that... just because my activity is different than yours doesn’t mean we are completely different.”

“Normal” for young adults with CF was comprised of variants related to their livelihood. For some participants, it embodied the ability to maintain their healthy appearance to the general public. To other young adults interviewed, “normal” was the experience to live with no symptoms requiring treatment. Participants described their normal as unique, and yet similar to their peers. For some young adult participants, “normal” meant fitting in and not standing out because of their disease.

It is important to understand the individual nuances of “normal,” as described by these participants. Every young adult participant mentioned their perception of “normal” and described the meaning. Some definitions were similar, related to health and appearances, while several participants felt that living with CF was their “normal.” One word – “normal” – encompassed internal and external experiences of young adults with cystic fibrosis.

CHAPTER 5

Discussion and Implications

The purpose of this study is to explore the impact of social isolation among young adults living with CF, and the findings identified the importance of *social connection and isolation*. The necessity for isolation from other individuals with CF is derived from the recommendation of the infection prevention and control guidelines. This recommendation piqued the researcher's interest in the experience of isolation among CF individuals, particularly among young adults with CF. The researcher found, however, that there was limited research that explored this experience. The primary findings revealed two main concepts: *social connection and isolation* and *defining "normal."*

Discussion

This exploratory research examines the experiences of ten young adults living with CF. The interviews elicited information pertaining to the participants' perceptions, experiences and understanding of *social connection and isolation*, to explore the impact of CF within various aspects of the participants' lives. The research findings identified challenges related to the impact of *social connection and isolation* on their disease management, friendships, and overall lifestyle. The theme of striving for normalcy emerged from several participants. The personal accounts of their lives and examples demonstrated their authenticity, vulnerability, and courage of individuals living with CF.

It is important to recognize the evolution of CF based on advances in medical technology and treatment of the disease. Historically, CF was seen as a fatal childhood disease. The Cystic Fibrosis Foundation, alongside medical clinicians and researchers, has worked tirelessly to increase the general public's understanding of the disease and the life expectancy for individuals with CF (Cystic Fibrosis Foundation Patient Registry, 2014). The average age of death of an individual diagnosed with CF in the United States is now 29 years (Cystic Fibrosis Foundation

Patient Registry, 2014), which was an important factor to understand when interviewing the young adult participants. The participants' knowledge of their disease has significantly impacted their experience of *social connection and isolation*, as many of them realize that they may only have a few years left to live.

Importance of Connection

A primary concept that emerged from the data was the *importance of connection*. Participants explored this theme supported by several pieces of the discussion, including their *perception of social networks*, conscious use of *social media*, degree of adherence to *infection prevention and control guidelines*, understanding dynamics of their *siblings*, their journey to determining *the value of friendships and supported autonomy* striving for independence while relying on their parents and significant others.

Infection prevention and control guidelines.

The *infection prevention and control guidelines* impact young adults' perception, understanding and experience of *social connection and isolation*. The researcher expected the guidelines would have led young adult participants to feel a greater sense of isolation and disconnection. Young adults with CF have unique relationships with their chosen support system and their environments. Young adults expressed challenges describing their health and daily concerns with their support system due to the lack of understanding and desire not to burden others with their disease. Their support networks provide care and support, however, it is not a shared experience. Several participants described feeling connected to their support systems; other participants discussed feeling isolated, unable to recognize that they were surrounded by a support network they created for themselves comprised of family, significant others, friends and their CF medical team.

The *infection prevention and control guidelines* impacted participants' ability to participate in face to face communication with other individuals living with CF. Throughout the

interviews, young adult participants expressed their frustrations with this recommended precaution. Some participants adhered to guidelines by avoiding face to face interaction with other CF individuals, while other participants disregarded this restriction – some quite deliberately. When asked how they perceive and experience social isolation, many participants denied immediate impact. Participants described scenarios where they felt disconnected, not necessarily from other individuals with CF, but from the general public because of the perceived stigma of their disease.

Siblings.

Siblings with CF within a single nuclear family are a unique and complex population. The study did not intend to explore sibling relationships, however, this material emerged from the data through inquiry about the infection guidelines. Several participants described the unique relationships with their siblings who were also diagnosed with CF compared to other participants' discussions of their siblings not diagnosed with CF. Young adult participants described the relationship with siblings also diagnosed with CF, by sharing their experience growing up being treated similarly. Siblings with CF are challenged by observing the different experiences of disease progression and disease management, which may be different from their own. This may include supporting their sibling while their disease is worsening or while they are being non-adherent with prescribed treatment regimens.

Given the genetic nature of the disease, there is often more than one sibling or family member diagnosed with CF. The findings from young adults with siblings also diagnosed with CF provide insight into health declines in young adults with CF based on differing disease management techniques. Multiple siblings with CF within the same family are a risk factor for poorer health outcomes (Lavie et al., 2014). Contact precautions related to the *Infection Prevention and Control Guidelines* have a different meaning for siblings diagnosed with CF. For example, being separated from each other by 6 feet does not apply. Siblings living with CF grow

up in the same household with exposure to the same pathogens. Once a CF sibling grows bacteria, it is likely their sibling will inevitably be infected. As such, younger siblings with CF tend to be colonized earlier with difficult to treat pathogens (Lavie et al., 2014). Once an older sibling has developed *supported autonomy* and moved out on their own, the overall health of the CF sibling that remains at home increases (Lavie et al., 2014). However, as this study's findings also demonstrate, siblings with CF develop a deep connection and understanding of the impact of the disease. The differences described by participants between themselves and their CF siblings typically focused on their acceptance of their disease and their adherence to disease management strategies. Young adult participants learned from their own experiences and from those of their siblings.

Young adults living with siblings not diagnosed with CF describe a different experience. These non-CF sibling relationships were described as being riddled with jealousy and misunderstanding. CF was perceived as a burden by participants, hindering their livelihood and negatively impacting their families. The non-CF sibling may observe the extra attention paid to their CF sibling related to intense daily treatments and occasional hospitalizations. The perception of the young adult participants related to their non-CF siblings described that their siblings not living with CF are aware of the attention paid to their CF sibling, causing them to feel less accepted than their CF sibling. There are few studies exploring the relationship between individuals living with CF and siblings. The perception described by these participants provides a counterpoint to the results from a sibling study, which utilized quantitative methods to report higher quality of life in siblings of individuals with CF when compared to peers with healthy siblings (Havermans et al., 2010). Older non-CF siblings reported greater impact of the disease than younger non-CF siblings (Havermans et al., 2010). This may lead to older siblings feeling more responsible and having an increased perception of disease burden. Siblings not diagnosed with CF may have a broader perspective, feel healthier, understand challenges and have an

increased ability to cope differently than their peers with health siblings (Havermans, et. al., 2010). However, the findings suggest that individuals with CF and their non-CF siblings may view the disease and its impact on family life quite differently.

Social media.

Young adults with CF have varying perceptions on their level of connection to CF and non-CF peers. *Social media* presents itself as a strength for young adults living with CF, providing an outlet to connect with others living with CF through Facebook, Instagram and other social networking platforms. Some young adults elect not to connect due to self-preservation or negative prior experiences. The ability to connect online provides individuals living with CF the opportunity to interact or engage with others that have a shared diagnosis. Communicating through online forums may help young adults cultivate connection and ameliorate feelings of isolation experienced by some CF patients, due to the *infection prevention and control guidelines* (Saiman et al, 2014). The current generation of young adults with cystic fibrosis understand technology, yet some participants described feeling uncomfortable texting or emailing strangers despite the commonality of living with CF. Online support forums (Johnson et al., 2001) lack the human connection a person receives through face to face interaction. Some young adults with CF have volunteered to provide peer support or mentoring through their CF centers, however, this opportunity is underutilized due to physical disconnect, lack of time and fear of the unknown.

Smoking and social seclusion.

The participants described their health as being threatened by others from the frequency of smoking in public. Smoke exposure was a significant concern for young adults with CF, as noted by participants and supported by the Cystic Fibrosis Foundation data. “In 2014, 22.7 percent of individuals with CF reported monthly or more frequent exposure to tobacco smoke, either secondhand or as a smoker themselves” (Cystic Fibrosis Foundation Patient Registry, 2014, p.29). Young adult participants in this study did not personally report their smoking habits,

however, they discussed the detrimental impact from others smoking in their surroundings.

Participants associated the experience of second hand smoke with increased feelings of seclusion from their friends, family and general public.

Value of friendships.

The importance of being accepted and understood by friends was a recurrent iteration by the young adults interviewed. They expressed the *value of friendships* and lessons learned regarding identifying the importance of real friends. Adherence to prescribed treatment is imperative for young adults with CF to maintain their health. Throughout their development, young adult participants described instances of avoiding treatments in order to spend more time with friends – this created a delicate balance between maintaining their health and having fun. Understanding the significance of quality relationships with others increased participants' feelings of connection.

CF is a unique chronic disease in the pediatric population because of the high treatment burden required to maintain good health. Other pediatric chronic diseases usually require one mode of prescribed therapy, such as inhaled medicines for asthma or insulin dosing for diabetes. In contrast, CF requires multiple modes of therapy on a daily basis, including inhaled medicines, chest physiotherapy, dietary changes and pancreatic enzyme replacement (Elgudin et al., 2004; Quittner et al., 2012). Similar to CF, adherence to prescribed treatment has been associated with improved health outcomes for individuals living with another rare chronic illness (Doyle & Werner-Lin, 2014). Cystic fibrosis is an invisible illness; even when individuals are feeling healthy, they are still required to perform daily treatment regimens to maintain their health. Some participants improved adherence when their friends were supportive and kept them accountable. Other participants described situations where they wanted to develop connections with others, and as such, they would avoid telling their peers about their disease and skip treatments to

establish friendships. Several young adults have learned through personal experience the value of treatment adherence and developing robust friendships.

Anxiety and depression.

Young adult participants reflected upon their own symptoms of *anxiety and depression*, which they were often reluctant to discuss with their CF medical teams. Young adult participants mentioned their long-term relationship with their CF team from their age at diagnosis through young adulthood. Participants discussed having a close relationship with their CF team, and subsequently did not want to divulge their feelings of anxiety and depression. They described seeking professional assistance outside of their CF care team. Participants described symptoms such as decreased feelings of connection and an increased perception of isolation. One female participant's story of suicidal ideation during high school and the following negative counseling experience is, unfortunately, not an isolated incident. It is imperative to recognize the severity of depression symptoms and understand, as mental health practitioners, the impact chronic illness has on the daily lives of young adults living with CF. According to the Cystic Fibrosis Foundation, prevalence of physical and mental health symptoms "is highest in early adulthood at a time when lung disease often worsens" (Cystic Fibrosis Foundation Patient Registry, 2014, p.68). Consistently conducting depression and anxiety screening is an important intervention when working with individuals living with CF.

The CFF acknowledges addressing mental health as a critical component of maintaining physical health and quality of life of individuals living with CF. The American Academy of Pediatrics (2014) recommends pediatric health care providers screen individuals over the age of 11 at primary care appointments. The aforementioned TIDES study results led to the development of CF specific anxiety and depression screening guidelines (Duff et al., 2014; Quittner et al., 2015). When the CFF developed and implemented the screening guidelines, it recommended that all accredited pediatric CF medical centers screen individuals diagnosed with

CF aged 12 and up (Quittner et al., 2015). Young adult participants expressed their understanding of the interconnectivity of mental and physical health on their disease management and outcomes.

Supported autonomy.

Young adults are yearning for independence and transformation to adulthood, however, they are uncertain of the complexities that accompany navigating this new stage of life. *Supported autonomy* emerged from the data through discussions of reliance on immediate family members and significant others for support. *Supported autonomy* is the relationship developed during this transformative time, between young adults and their family or significant others, whom they look to for continued support. Young adults rely on their family or significant other for several disease management tasks, motivation for daily treatments, accompaniment to doctor appointments, visitation during hospitalization, and above all, the feeling of being connected. These young adults on their journey to adulthood conveyed more responsibility, disease understanding and treatment burden. Young adults with CF strive to balance their social life with disease management. Many accept help from immediate family members and significant others to provide physical and emotional support. Other young adults, however, struggle to reach out and ask for help as a self-preservation technique to rely solely on their independence and personal perseverance.

Emerging adulthood, a concept pioneered by Arnett (2000), is a newly-defined developmental period that captures the experience of the transition common in Western, industrialized, and resource rich areas. The phrase was coined to describe the time when individuals no longer view themselves as teenagers but also do not perceive themselves as fully capable or functioning adults (Arnett, 2000). Chronic disease processes complicate the movement into adulthood, and individuals with chronic illness may not rely on traditional milestones such as finishing their education or getting married to define adulthood (Doyle &

Werner-Lin, 2014). Young adults living with cystinosis, another rare genetic disease, described themselves as incomplete adults as they transitioned to adulthood and adult oriented care (Doyle & Werner-Lin, 2014). Young adults with CF find themselves amidst life changes and challenges, including typical developmental milestones, in addition to increasing responsibilities for their disease management.

This study echoes the findings presented by the Cystic Fibrosis Foundation regarding young adults with CF pursuing higher education, employment, and committed relationships (Cystic Fibrosis Foundation Patient Registry, 2014). Many young adults between the ages of 18 and 25 living with CF lead active lives: they go to college, pursue careers, enter romantic relationships and marriages. Despite this, young adults with chronic illness still frequently rely on their families of origin for emotional and financial support (Doyle & Werner-Lin, 2014). Young adults with CF develop *supported autonomy* and often share the burden with their family or significant other. The *importance of connection*, related to the impact of *social connection and isolation* among young adult participants with CF, remains key to the ongoing health and well-being of these young adults as they maintain and renegotiate reliance on their family of origin.

Defining “Normal”

The discussion of “normal” emerged from all ten young adult participants, as they described what living normally meant to them. Although this topic was not part of the original interview guide, the researcher identified the inherent need to explore further, thus, the subsequent concept *defining “normal”* developed. Participants’ attached their own meaning to the word “normal,” given their understanding and perception of their disease and impact it has on their daily lives. Some participants were diagnosed in infancy, and as such, this is the only life they have ever known – this is “normal” for them. Among participants, regardless of their age at diagnosis, some have accepted CF and expressed that their lives were “normal.” Other participants were striving for this notion of normalcy, as compared to their non-CF peers. Young

adults explored being accepted and understood by others in general, and “seeking normalcy” (Schulman-Green et al., 2012, p. 141) in chronic illness is supported by the participants’ discussion of balancing their treatments with daily lives.

Direct Practice Implications

Recognizing the *importance of connection* may aid health care providers and social workers on a multidisciplinary team to understand their young adult CF patients. The Cystic Fibrosis Foundation recommends social workers evaluate their patients at least once a year (Cystic Fibrosis Foundation Patient Registry, 2014). Utilizing systems theory and a biopsychosocial approach allows CF social workers to conduct annual assessments which go beyond the patient’s disease, in order to understand the young adult’s experience of the illness and to develop meaningful interventions. The social worker may assist the young adult patient to understand their right to self-determination, an ability to autonomously make well informed decisions regarding their care. CF social workers have several roles that include: performing biopsychosocial assessments, discussing and promoting treatment adherence, screening for anxiety and depression, and providing resources and referrals. Yet, there is a paucity of knowledge related to understanding the impact of *social connection and isolation* on disease management and how young adults are *defining “normal”* within their lives and illness experience. Increased understanding of their perception of “normal” and their experiences of *social connection and isolation*, in addition to the recognition of CF’s impact on mental health, will likely help social workers and the CF medical care team formulate specific treatment plans to improve health outcomes.

Expanding assessment beyond patient’s daily routine and regimen can allow social workers to explore barriers to self-care management and adherence more deeply and can provide insightful information regarding the young adults’ social lives and the nuances of their disease experiences. CF social workers can assist young adults living with CF to feel less isolated

through identifying ways to balance treatment adherence with participation in social activities with their peers. This will lead to improved quality of life and better outcomes. Social workers can assist young adult patients in taking ownership of their disease management, and accepting their “normal” identity of living with CF. In order to achieve these goals, social workers will need to elicit further information than is currently ascertained on routine CF social work evaluations. Additional questions to add to social worker’s annual biopsychosocial assessment regarding perceptions of disease management and socialization include:

- 1) What is a typical day like for you?
- 2) What about having CF do you find challenging?
- 3) How does living with CF affect your social life?
- 4) Are you satisfied with the time you spend socializing with you family or peers compared to the time you spend conducting your treatment?
- 5) Do you know anyone living with CF? If yes, are there ways in which you communicate with this person?
- 6) Would you like to connect to or communicate with other people living with CF?
- 7) Do you use social media? If yes, which avenues of social media do you utilize?
- 8) Who are the individuals in your social network?
- 9) What is normal for you?
- 10) If you could change anything what would it be and why?

Briefly review the infection prevention and control guidelines with the young adult and then inquire how this impacts them. These questions may open communication between the social worker and patient to discuss connection to others and bridge the gap between *social connection*

and isolation. Social workers need to be comfortable in exploring these difficult topics such as anxiety and depression, challenges to living with the disease, impact of infection prevention and control guidelines, asking about the impact of CF on their social connections, and having a better understanding of the young adult patient's perception of normal. Strong rapport between the patient and social worker is vital to explore these types of topics in a way that ensures the patient is being forthcoming and comfortable sharing their true experience. These conversations may increase the young adults' awareness of how these areas impact their daily life, improve outcomes and bridge the gap of social isolation. Asking these questions will open dialogue allowing the social worker to develop deeper understanding of the young adults meaning, perception and experiences of *social connection and isolation* along with their definition of "normal." This will also enhance the social worker's level of connection with the patient and awareness of disease acceptance.

Young adult CF patients have unique experiences, understandings and perceptions that share a common theme: they are all living with CF and knowledge of the average life expectancy. Through having a strong patient social worker relationship, social workers can empower young adults to advocate for themselves and explore ways in which they can better balance treatment adherence, socializing with peers, and living the life they perceive to be "normal." Engaging in problem solving can allow for the development of individualized treatment plans and meaningful patient specific interventions. Young adults with CF described being screened for anxiety and depression, however, are hesitant to discuss their symptoms. Social workers and CF medical teams need to be cognizant when screening for anxiety and depression that patients are not always forthcoming with their symptoms during screening. The social workers need to ensure the patient can openly communicate their symptoms, concerns, and disease management techniques to their CF health care team.

Social workers maneuver the delicate balance between medical recommendations, physician expectations, available resources, and the patient's needs and preferences. The additional assessment questions allow the social worker to gain insight and better understanding of the patient's perceptions leading to the development of patient specific interventions. While treatment plans may consist of similar elements and techniques, individualized interventions can be developed within the social worker and patient dyad, based on patient's preferences and beliefs, in order to achieve patient goals. Meaningful interventions may include connecting the patient with identified resources, peer mentoring, virtual support through social media, connecting their significant others and families to caregiver support groups, assisting siblings with understanding the nuances of the chronic illness, among other variations of treatment plans. It is essential to incorporate the patient into treatment planning, and remember the patient is the primary decision-maker in their own care. From a strengths-based perspective, social workers in direct practice with patients and families can support the value of dignity and worth of a person (National Association of Social Workers, 2008), while respecting their CF patient's right to self-determination.

This research highlights the impact of *social connection and isolation*, providing an opportunity for social workers and the health care team to incorporate conversations into the treatment of young adults with CF and other chronic illnesses. The dialogue promotes better understanding and awareness of how *social connection and isolation* effect the young adults' lives and disease management. Social workers will gain deeper understanding into the perspective and experiences of young adults living with CF and other chronic illnesses. The assessments and development of meaningful interventions will engage young adults in their feelings around *social connection and isolation*, additionally gaining deeper insight into the young adults' psychological and social development.

Implications for Future Research

The social implications of CF are changing as the lifespan of individuals living with CF increases. The years of contributions by researchers and advocates have improved disease understanding and outcomes. There continues to be CF research performed related to genotyping, bacteria, medications, and airway clearance, however, research concerning psychosocial and mental health impacts is not yet included as a core component of the CF literature. While childhood impact has been extensively studied, a gap remains in the literature regarding the impact of living with this disease in emerging adulthood and beyond. Future research exploring the relationship between individuals with CF and their CF and non-CF siblings could provide unique insight into the impact of CF on family function and structure. Further research related to *social connection and isolation* during young adulthood for those with other chronic and rare diseases is also warranted.

Limitations

Limitations to this study include the small sample size of ten participants. The participants interviewed in this study were primarily female; the study was comprised of nine young adult females and one male. Additionally, those who participate in research are more likely to have positive coping strategies. The findings describe sibling relationships although there were no siblings interviewed in this study. All of the participants were Caucasian, which represents the majority of individuals with CF living in the United States. It is important to note that the experience of young adults with CF may be different outside of the United States. Additional research is warranted to further explore the implications of *social connection and isolation* among a broader sample of young adults with CF.

Conclusion

This study sought to explore the impact of social isolation of young adults living with CF, particularly in light of infection control guidelines that recommend avoiding in-person contact

with others who share their experience living with the disease. The participants described their desire for connection, as they sought to be understood and perceived as “normal.” In spite of their disease and treatment burden, young adults with cystic fibrosis are self-aware and develop connections to their support networks. Young adult participants with CF demonstrated their ability for social connection and developed techniques for reducing isolation through social networks, social media, sibling relationships, and friendships. They discussed their personal journey living with CF, described their experiences, and explored their perceptions and understanding of the *importance of connection* while sharing insight for their future and the future of those living with the disease.

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Appendix A. IRB Approval

University of Pennsylvania
Office of Regulatory Affairs
3624 Market St., Suite 301 S
Philadelphia, PA 19104-6006
Ph: 215-573-2540/ Fax: 215-573-9438
INSTITUTIONAL REVIEW BOARD
(Federalwide Assurance # 00004028)

09-Sep-2015

Allison V Werner-Lin
Attn: Chelsea Toth
ctoht@sp2.upenn.edu
awerl@sp2.upenn.edu

PRINCIPAL INVESTIGATOR : Allison V Werner-Lin
TITLE : Cystic Fibrosis and Social Isolation During Young Adulthood
SPONSORING AGENCY : No Sponsor Number
PROTOCOL # : 823068
REVIEW BOARD : IRB #8

Dear Dr. Werner-Lin:

The above-referenced research proposal was reviewed by the Institutional Review Board (IRB) on 9/8/2015. It has been determined that the proposal meets eligibility criteria for IRB review exemption authorized by 45 CFR 46.101, category 2.

This does not necessarily constitute authorization to initiate the conduct of a human subject research study. You are responsible for assuring other relevant committee approvals.

Consistent with the federal regulations, ongoing oversight of this proposal is not required. No continuing reviews will be required for this proposal. The proposal can proceed as approved by the IRB. This decision will not affect any funding of your proposal.

Please Note: The IRB must be kept apprised of any and all changes in the research that may have an impact on the IRB review mechanism needed for a specific proposal. You are required to notify the IRB if any changes are proposed in the study that might alter its IRB exempt status or HIPAA compliance status. New procedures that may have an impact on the risk-to-benefit ratio cannot be initiated until Committee approval has been given.

If your study is funded by an external agency, please retain this letter as documentation of the IRB's determination regarding your proposal.

Please Note: You are responsible for assuring and maintaining other relevant committee approvals.

If you have any questions about the information in this letter, please contact the IRB administrative staff. Contact information is available at our website: <http://www.upenn.edu/IRB/directory>.

Thank you for your cooperation.

Sincerely,

**Benjamin
Hernberg**

IRB Administrator

Digitally signed by Benjamin
Hernberg
DN: cn=Benjamin Hernberg, o=IRB,
ou=IRB, email=benhernberg@upenn.edu,
c=US
Reason: I am the issuer and
integrity of this document
Date: 2015.09.08 14:09:19 -0400

Appendix B. Recruitment Flyer

Living with Cystic Fibrosis in Early Adulthood

University of Pennsylvania
School of Social Policy and Practice
Chelsea Toth, MSW

Who is Eligible?

- ❖ Males and Females with CF 18 – 25 Years Old
- ❖ Ability to Read and Speak English
- ❖ Not Currently Hospitalized
- ❖ Only One Sibling From Any Family

I'm interested in researching the impact of the prevention infection and control guidelines on young adults living with CF in their early adult years.

The voluntary interview will last about an hour. Each participant will receive a \$15 Amazon gift.

This study is part of dissertation research for Doctorate in Clinical Social Work from the University of Pennsylvania. The project has been approved by the University of Pennsylvania Institutional Review Board.

For questions or concerns, please contact Chelsea Toth

732.616.9370 ctoth@sp2.upenn.edu

Appendix C. Recruitment Email to Cystic Fibrosis Foundation

Email Subject: Young Adults with CF Study

To Whom It May Concern:

As a CF social worker, I understand the importance of allowing our patients to express themselves. I am currently working on a research project as part of the completion of my Doctorate in Clinical Social Work. My research is focused on the impact of CF during the transition to adulthood.

I am inviting young adults with CF between the ages of 18 and 25 who speak English to discuss their experiences with CF. I am looking to interview young adults with CF during the summer and fall of 2015. The voluntary interview will last approximately 60 minutes. Each participants will receive a \$15 Amazon gift card.

The results of this project will be used for my dissertation at University of Pennsylvania School of Social Policy & Practice. I expect to use the data for my dissertation, which will be available to you upon completion.

I have attached a flyer to this email. I would greatly appreciate if you could pass along this information to the CF community, including posting the flyer on the CFF Facebook page. If you have any questions or concerns, please feel free to contact me at 732-616-9370 or via email ctoth@sp2.upenn.edu.

This project has been approved by the University of Pennsylvania Institutional Review Board.

Thank you in advance!

Sincerely,

Chelsea Toth, MSW
DSW Candidate
School of Social Policy and Practice
University of Pennsylvania

Appendix D. Recruitment Email to CF Social Workers

Email Subject: Young Adults with CF Study

CF Social Workers:

As a CF social worker, I understand the importance of allowing our patients to express themselves. I am currently working on a research project as part of the completion of my Doctorate in Clinical Social Work. My research is focused on the impact of CF during the transition to adulthood.

I am inviting young adults with CF between the ages of 18 and 25 who speak English to discuss their experiences with CF. I am looking to interview young adults with CF during the summer and fall of 2015. The voluntary interview will last approximately 60 minutes. Each participants will receive a \$15 Amazon gift card.

The results of this project will be used for my dissertation at University of Pennsylvania School of Social Policy & Practice. I expect to use the data for my dissertation, which will be available to you upon completion.

I have attached a flyer to this email. I would greatly appreciate if you could pass along this information to your young adult patients. If you or your patients have any questions or concerns about participating in this study, please feel free to contact me at 732-616-9370 or via email ctoth@sp2.upenn.edu.

This project has been approved by the University of Pennsylvania Institutional Review Board.

Thank you in advance!

Sincerely,

Chelsea Toth, MSW
DSW Candidate
School of Social Policy and Practice
University of Pennsylvania

Appendix E. Consent Form

Terms of Informed Consent and Study Requirements:

Purpose/Procedure: You are being asked voluntarily to participate in this study because you are 18-25 years old and have been diagnosed with cystic fibrosis. The purpose of the study is to learn more about the perceptions of social isolation and the impact it has during your transition to adulthood. This study is being conducted for a dissertation in social work. Participation in this study involves completion of a one-time interview, which will last approximately 60 minutes. This study is being conducted through the University of Pennsylvania. However, the interview is online and can be accessed on any computer.

Risks: There are minimal risks involved in participating in this study. Risks may include feeling uncomfortable or embarrassed with sharing your opinions and beliefs or meeting the host and being audio recorded during the session. You also will be giving up approximately 60 minutes of your time to participate in the study; therefore, this may impact your personal responsibilities. However, should you find the questions upsetting for any reason, you may discontinue the survey at any time.

Benefits: There is no benefit to you. However, your participation could help us understand the perception experiences of young adults with CF, which may benefit you indirectly. In the future, this may help other people to better assist young adults with CF.

Confidentiality: Every effort will be made to keep information obtained during this study confidential. We will keep any records that we produce private to the extent we are required to do so by law. The records of this study will be kept confidential and any information collected through this research project that personally identifies you will not be voluntarily released or disclosed without your separate consent, except as specifically required by law. In any sort of report we might publish, we will not include any information that will make it possible to identify a subject. Research records will be stored securely and only researchers will have access to the records. The interviews will be audio recorded; however, the names and other identifying information will not be used while recording, as to keep your identity confidential. Only the researchers will have access to these audiotapes, and tapes will be erased from devices and deleted from computer files at the conclusion of this study. However, we cannot guarantee total privacy. Records can be opened by court order or produced in response to a subpoena or a request for production of documents. If information from this study is published or presented at scientific meetings, your name and other personal information will not be used.

Voluntary Participation and Withdrawal: You must be 18 years of age or over to participate in this study. Participation in this study is voluntary; you are free to refuse to answer any questions, or to withdraw at any time. Each participant is free to withdraw consent and discontinue participation in the study at any time.

Reward: Participants will each receive a \$15 Amazon gift card at the completion of the interview.

Contact Persons: If you have questions, concerns or complaints regarding your participation in this research study or if you have any questions about your rights as a research subject, you should speak with the Student Investigator, Chelsea Toth at ctoth@sp2.upenn.edu. If a member of the research team cannot be reached or you want to talk to someone other than those working

on the study, you may contact the Office of Regulatory Affairs with any question, concerns or complaints at the University of Pennsylvania by calling (215) 898-2614.

You will be given a copy of this information to keep for your records.

By checking this box, I am agreeing that I am 18 years of age or over and am agreeing to the terms of the informed consent and study requirements.

- I agree that I am 18 years of age or over and am agreeing to the terms of the informed consent and study requirements.

Appendix F. Demographic Questionnaire

Age: _____

Are You

- _____ Male
- _____ Female

Marital Status

- _____ Single
- _____ Dating
- _____ Married/Partnered
- _____ Separated/Divorced
- _____ Widowed

At what age were you diagnosed with cystic fibrosis?

- _____ Age Years
- _____ Age Months

What do you call your disease?

Have you ever received a transplant?

- _____ Yes
- _____ No

Highest level of education completed?

- _____ Some High School
- _____ High School Diploma
- _____ Some College
- _____ Associate's Degree
- _____ Bachelor's Degree
- _____ Graduate/Professional Degree
- _____ Other: _____

Employment Status:

- _____ Working Part-time
- _____ Working Full-time
- _____ Not working for other reasons
- _____ Not working due to health reasons
- _____ Other: _____

Insurance Coverage (select all that apply)

- _____ Private
- _____ Medicaid
- _____ Medicare
- _____ Tricare
- _____ Affordable Care Act

What is your ethnicity?

- _____ Caucasian
- _____ African American
- _____ Asian
- _____ Hispanic
- _____ Native American
- _____ Pacific Islander
- _____ Other: _____

Appendix G. Interview Guide

Semi-Structured Interview Questions

The study will explore the ways experiences living with cystic fibrosis impact your development during the young adult years.

Research Questions: This study aims to explore three primary and related research questions: How do individuals with CF aged 18-25 understand, experience, and perceive social isolation from others with CF? Are there others ways in which young adults with CF experience isolation? How does social isolation impact their medical and psychosocial development during the young adult years?

Living with CF

- Tell me about living with CF
- What is a typical day like for you now?
- Tell me about the ways in which CF has impacted your daily life? Your relationships with family, friends, romantic partnerships?
- A lot of people have talked about having a hard time with _____, can you tell me about your experience..
- If you were talking to someone who didn't know anything about CF, how would you describe it? What you want them to know?

Probe

- Do you know your genotyping? How does this knowledge affect your experience?
- Tell me more about the situation you mentioned...
 - Can you clarify _____ or give an example?

Infection Prevention and Control Guidelines

- What do you know about the Infection Prevention and Control Guidelines?
- Are there ways in which the guidelines impact your daily life? Tell me about these.
- The guidelines suggest limiting face to face contact from other individuals with CF, how does this impact your daily life? - - do you have any siblings?

Probe

- In what ways does this affect you at medical appointments, school, camp, advocacy events?

Social Isolation

- What has been the most challenging part of living with CF?
- Do you interact or communicate with other people with have CF? How?
- Tell me about a situation that made you feel isolated from others with CF?
- What are some avenues you use to feel less isolated?

Probe

- How have you reacted/responded to _____ feelings?
- Can you explain _____, in greater detail?

Adolescence

- In your experience, how have decisions made during adolescence regarding treatment impacted your current health?
- What would you recommend to adolescents with CF?

Probe

- If you could change something you did during adolescence related to CF what would it be?
- If you could tell your teenage self, something – what would it be?
- Your experience is very interesting, can you return to earlier when you mentioned...

Future

- Is there anything about your life with CF you would like to add that I haven't asked you?
- Where do you see yourself in five years?
- What do you want to tell others with CF?
- Thoughts about the interview?

Appendix H. Code Book

THEME	CODES	DEFINITION
CF Diagnosis	<i>CF Definition</i>	<p>Perceived scientific definition discussed by participants. CF is a genetic disease that affects the lungs and digestive system. Disease produces thick sticky mucus. Progressive disease individuals cough often which is not contagious. Participants discussed pancreatic insufficiency requiring pancreatic enzymes before all meals and snacks to absorb nutrients. Negative effects of not taking enzymes include upset stomach and increased need to use the bathroom.</p> <p>Ex: start with very scientific definition, “genetic disease that mainly affects the lungs”; “Thick sticky mucus in my lungs often makes it difficult to breathe”; “it’s not like you can catch CF”; Ex: friends unaware of enzymes, “gotten good at it”</p>
	<i>Impact of Age at Diagnosis</i>	<p>Varying degree of importance at age of diagnosis. Participants explored awareness of CF from infancy versus being diagnosed later in childhood/adolescence.</p> <p>Ex: diagnosis turned world upside down; diagnosed at 12 and changed my life; remember the day I was diagnosed and both parents were there; “I didn’t know what CF was at all”; “for the first time in months I could breathe”; deeper appreciation for life; luckily diagnosed really early; diagnosed during infancy – grew up entire life knowing had CF; “I don’t wish I was diagnosed earlier”...liked having years of blissful ignorance.</p>
	<i>Information Seeking</i>	<p>Lack of personal understanding about disease severity, conducted research to learn about the disease. Translates to insufficient information in the general public’s unawareness of CF, asking a lot of questions and exhibit pity looks.</p> <p>Ex: “then I say, but I’m healthy”</p>
	<i>Genotyping</i>	<p>Participant’s awareness of genotyping and gene specific mutation important knowledge with advances in medical technology including gene specific medications.</p> <p>Ex: lung function improved on Orkambi; since on Orkambi lung function doubled; disappointed doesn’t qualify for genotyping “just waiting”; “I got both CF genes”; “I was doubtful that this first one that hits the market is going to be super impactful for me”</p>

THEME	CODES	DEFINITION
<p>Degree of Familial Support</p>	<p><i>Parental Influence</i></p>	<p>Participants discussed impact of positive parental caregiving on family cohesiveness versus lack of parental/familial acceptance led to negative health outcomes.</p> <p>Ex: good support system impactful and makes a difference. CF changed relationship with mother, has not been as close to her, “she treats it like CF is my life...I don’t think she can get away from that umber about our life expectancy”; “sitting in the bathroom with my mom with the shower running on hot to try to get some moisture in my lungs”</p>
	<p><i>Financial Burden</i></p>	<p>Perceived financial impact due to cost of medications, hospitalizations and insurance denials. Emotional distress on parents and children.</p> <p>Ex: Mother increased hours for financial stability. One Christmas Mother told me my present that year was going into the hospital. Got less gifts than non-CF siblings. Struggled to make ends meet with medications.</p>
	<p><i>Supported Autonomy</i></p>	<p>Developing individuality while remaining attached to their family and significant others. Individuals want decision making capacity and hesitant to ask for help. Meanwhile, parent or significant other attend appointments and visit during hospitalizations.</p> <p>Ex: problems communicating when actually sick. Hardest part is being open and allowing someone to be there through the thick and thin. Learning how to lean on people. “not that I’m living on my own and having to take care of myself it’s a little harder”; significant other: “knows how I want to live my life”; “a real life adult”; “he very much gets it and has been helpful” ; my mom keeps me accountable and can tell when I’m getting sick</p>
	<p><i>Accountability</i></p>	<p>Appreciate parents and significant other continued encouragement to adhere to medical treatments, physical activity and doctors’ appointments. Primary caregiver historically the enforcer and advocate with community members. Reflect upon primary caregiver leading disease management with the CF medical care team.</p> <p>Ex: Mom kept accountable as a teenager. “making sure we did our treatments”; “Mom always made sure that we stuck to our stuff”</p>

THEME	CODES	DEFINITION
Impact of Life Expectancy	<i>Preoccupation with Death</i>	<p>Knowledge about life expectancy, awareness of disease progression, utterances of death, impact of dying on loved ones left behind, being told going to die young led to life limiting perspective.</p> <p>Ex: “I spent my whole life thinking I was gonna die and here I am and I’m not dead”; “CF isn’t gonna take me one day. It’s a slow process”; built entire life on assumption “I would be dead before I got there”; “all my plans are centered around me dying” ; you can’t live your life like you’re gonna die tomorrow or when you’re 23, “if you focus so much on the thought that you’re dying then you stop living”; “Thought was dying”</p>
	<i>Mental Illness Symptoms</i>	<p>Participants explored topics surrounding anxiety, worry, depression and suicidal ideation.</p> <p>Ex: “the things that I have to worry about that no one else does”; worry about disease progression and worry CF is getting worse; “if you don’t physically feel well that can impact your mental health drastically”; “I feel like I don’t have enough time when I worry I am going to get sick”; “I’m just having trouble finding myself”; suicidal ideations thought it was OK if I didn’t want up the next day; “look back in disbelief mental health got that bad”;</p>
	<i>Future Planning</i>	<p>Optimism and skepticism about future goals, accomplishments and health.</p> <p>Ex: graduate college, get married, have children, buy a house and own animals; “worry am I going to be able to have kids”; “the longer I can be there to see my kids grow up the better”; “personally all I want to do right now is grow old with my husband and have three beautiful children”; “whatever CF does to me I have plans for my own life”; “would rather die from CF than give up those plans”</p>
	<i>Engaging in Mental Health Services</i>	<p>Weighing pros and cons of seeking professional mental health provider on long term impact of disease management.</p> <p>Ex: stigma with depression and diagnosis perceived a lot of people don’t want to touch it, saw unhelpful counselor in high school “told me all of my problems are centered around the fact that I have a chronic disease”; “I just need somebody to listen not judge me and not have a biased opinion”; recent therapist suggested an anti-depressant.</p>

THEME	CODES	DEFINITION
Prioritizing Health	<i>Hospitalization</i>	<p>Understanding the difference on health perception for scheduled “tune-up” hospitalizations versus unscheduled admission. Discussion of frequency in the hospital and treatment regimen during admission.</p> <p>Ex: “I’ve only been hospitalized one time in only 19 years, which is huge”; “thrown into the hospital without being prepared”; scheduled hospitalizations to be desensitized to new IV antibiotics; developed allergies to antibiotics; transition to adult hospital at 26 due to CF kids not living as long; hemoptysis “terrifying” experience led to four years in and out of the hospital.</p>
	<i>Importance of lung function</i>	<p>Utterance about knowing and monitoring their lung function. Includes reacting to FEV1 and overall lung function numbers.</p> <p>Ex: higher lung function than most normal people in general; at diagnosis PFTs 32, current PFTs 55, highest 95.; “I don’t consider myself an average CF patient” because of where my numbers are.; PFT average 90 dips down to 63 hospitalized; PFTs down quite a bit, interpreted doctor phrase “you’re getting older you might actually start having CF now”</p>
	<i>Turning point</i>	<p>Experiences related to serious health awareness evoking a level of fear, recognizing the need to become self-aware and change behaviors to take better care of their disease.</p> <p>Ex: Big panic when something out of the ordinary happens; “first time that CF really terrified me was the first time I coughed up blood”; “the moment I woke up and needed to know what was going on in my own body” paid attention to doctors, lung function and weight; being unhealthy not helping me or my family; flipped switch “determined to get healthier and stay healthy”; realize CF is not going to go away... didn’t do vest for an entire year got sick and tired of doing treatments and antibiotics...realized have to start takin care of myself and making health a priority.</p>
	<i>Infection Prevention and Control Guidelines</i>	<p>Explicit or implicit references to acknowledging the prevalence of the Infection Prevention and Control Guidelines. Include respect for medical staff following guidelines versus personal disregard for the recommendations.</p> <p>Ex: doctors gown and glove when enter exam room; understand wear a mask to protect from getting germs, annoying to wear mask; wear mask “that particular guideline was throwing every CF patient under the bus” ; aware of 6 foot rule don’t be in same space with other people with CF “be really cautious when you’re around other people with CF and avoid if possible”; “people with CF aren’t supposed to be within close proximity of each other for risk of passing lung infection or bacteria back and forth”; expressed frustrations with guideline restrictions “the people that understand you, you can’t spend time with”; “at the end of the day I like to have a face to face conversation with someone”</p>

THEME	CODES	DEFINITION
Reality of living with CF	<i>Concept of Adolescence</i>	<p>Participants discussed treatment adherence during adolescents led to perceived improved current health outcomes. Non-adherence to prescribed treatment during the rebellious adolescent phase led to feelings of remorse on negative health consequences.</p> <p>Ex: would be better if did treatment during adolescence and established a routine as a teenager; medicine helps even if you think it's not; tell myself whether in the mood to do treatment or not, always remember treatment and health come first; "the more damage you do to your body as you're younger is not going to help you in the long run"; "I did everything the doctor told me to do, if I hadn't done that, I would not be healthy"; Recommend to adolescents with CF: "do what your doctors tell you do to"; if you take the time to do what you need to do, you will end up spending less time on it in the long run"</p>
	<i>Treatment Regimen</i>	<p>Importance of establishing daily routine to include intensive time consuming prescribed airway clearance and medical treatments. Discussion of time management, challenging finding the balance and maintaining overall health and daily functioning.</p> <p>Ex: Treatment regimen: vest, albuterol, pulmozyme, cayston every other month; time consuming regimen; repeat next day; two hours of day dealing with CF; morning treatment routine "then I get ready like everybody else"; can't come out now need to do a treatment; some people do chores or other responsibilities my normal is getting up, go to work and do treatments; "always feel like I have to be active"</p>
	<i>Defining Normal</i>	<p>Perception of defining normal compared to non-CF peers related to not taking medication, fewer hospitalizations and less to worry about health. Many participants expressed feeling "normal" living with CF.</p> <p>Ex: definition of normal not having to get up and do treatments and take all medication; "a normal person doesn't have a lot of worries when it comes to health"; "I always refuse to wear the mask because I just like to feel normal"; "I actually think my life is pretty normal"; "basically living a normal life except we just have to do treatments at night and took pills when we ate"; "can't really work the way other people do"</p>
	<i>Individual Health</i>	<p>Utterances about health individuality, degree of severity and appearances.</p> <p>Ex: "You look at me and you have no idea that I have anything wrong with me"; "want them to know that even though I look healthy, it takes a lot of extra work to stay healthy"; "not to assume everybody who has CF is going through the exact same thing"</p>

THEME	CODES	DEFINITION
Importance of Connection	<i>Social Isolation</i>	<p>Utterances of frustrations not being understood, feeling connected and disconnected from others, aware of the impact their disease has on making connections. Participants weighed impact of isolation from other people in general and lacked feeling isolated from others with CF.</p> <p>Ex: felt isolated when friends don't understand; feel isolated from other people in general; couldn't go to sleepovers due to illness; no desire to meet anyone with CF; "I don't really feel isolated at all"; it never occurred to them that smoking outside could be harmful to somebody"; feeling isolated / judged "people don't take a moment to consider what I'm going through"; "maybe she's coughing because she has a serious illness"</p>
	<i>Siblings</i>	<p>Explicit and implicit differences and similarities expressed for CF sibling versus non-CF sibling relationship. Difference in disease management techniques and acceptance between CF siblings.</p> <p>Ex: all three siblings have CF and have been relatively healthy whole lives "my brother and sister also have CF"; CF sister health deteriorating adds stress and worry to relationship "hopefully she turns around"; sister wants to be CF doctor "since she was 7 years old my sister told me she was going to cure me" ... "I inspired my baby sister and that is so meaningful to me"; never realized impact of CF on non-CF sibling; CF is scary for non-CF sibling; non CF sibling feels left out and feels CF sibling is the favorite; sister thinks I'm perfect in her mind and can do no wrong "spends as much time with her so that she knows I love her"</p>
	<i>Value of Friendships</i>	<p>Two different types of friendship discussed positive supportive friends versus negative sympathy friends. Important for friends to understand intricacies of having CF.</p> <p>Ex: "felt special that real friends went out of their way to make sure that I was included"; "having CF has brought more friends into my life"; "I don't want sympathy friends"; "I've figured out how to weed those out and get the friends that actually care about me and not just my story"; "they'll be there for you if they're good friends and if they don't stick with you then they aren't good friends"; "don't waste my time with people who don't understand it or don't care or are kind of intolerant of it"</p>
	<i>Social Media</i>	<p>Utilize Facebook and social media to spread awareness, give and receive advice, support, and have understanding CF community.</p> <p>Ex: follow CF individuals on social media; feel comfortable with CF Facebook group; "I know what you're going through let's be friends on Facebook"; "If I were going to reach out to someone with CF it would be online or something never in person"; share high calorie recipes; don't want to interact with people who have CF right now</p>

Appendix I. CITI Program

COLLABORATIVE INSTITUTIONAL TRAINING INITIATIVE (CITI PROGRAM) COURSEWORK REQUIREMENTS REPORT*

* NOTE: Scores on this Requirements Report reflect quiz completions at the time all requirements for the course were met. See list below for details. See separate Transcript Report for more recent quiz scores, including those on optional (supplemental) course elements.

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 • **Institution Affiliation:** University of Pennsylvania (ID: 898)
 • **Phone:** 732-616-9370

• **Curriculum Group:** Human Research
 • **Course Learner Group:** Social/Behavioral Research Course
 • **Stage:** Stage 1 - Basic Course
 • **Description:** Choose this group to satisfy CITI training requirements for Investigators and staff Involved primarily in social/behavioral research with human subjects.

• **Report ID:** 14860671
 • **Completion Date:** 01/17/2015
 • **Expiration Date:** 01/16/2018
 • **Minimum Passing:** 80
 • **Reported Score*:** 99

REQUIRED AND ELECTIVE MODULES ONLY	DATE COMPLETED	SCORE
University of Pennsylvania (ID:1697)	01/01/15	No Quiz
Belmont Report and CITI Course Introduction (ID:1127)	01/01/15	3/3 (100%)
Students In Research (ID:1321)	01/01/15	10/10 (100%)
History and Ethical Principles - SBE (ID:490)	01/01/15	5/5 (100%)
Defining Research with Human Subjects - SBE (ID:491)	01/01/15	5/5 (100%)
The Federal Regulations - SBE (ID:502)	01/01/15	5/5 (100%)
Assessing Risk - SBE (ID:503)	01/01/15	5/5 (100%)
Informed Consent - SBE (ID:504)	01/01/15	5/5 (100%)
Privacy and Confidentiality - SBE (ID:505)	01/01/15	5/5 (100%)
Research with Prisoners - SBE (ID:506)	01/16/15	5/5 (100%)
Research with Children - SBE (ID:507)	01/16/15	5/5 (100%)
Research in Public Elementary and Secondary Schools - SBE (ID:508)	01/17/15	5/5 (100%)
International Research - SBE (ID:509)	01/17/15	5/5 (100%)
Internet-Based Research - SBE (ID:510)	01/17/15	5/5 (100%)
Research and HIPAA Privacy Protections (ID:14)	01/17/15	5/5 (100%)
Vulnerable Subjects - Research Involving Workers/Employees (ID:483)	01/17/15	4/4 (100%)
Conflicts of Interest in Research Involving Human Subjects (ID:488)	01/17/15	4/5 (80%)

For this Report to be valid, the learner Identified above must have had a valid affiliation with the CITI Program subscribing Institution Identified above or have been a paid Independent Learner.

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