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Checkmate: Exploring Father-Son Communication Regarding Reproduction and Sexual Health in Males With Cystic Fibrosis

Dianna C. Sanderson

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Checkmate: Exploring father-son communication regarding reproduction and sexual health in males with cystic fibrosis

by

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I am grateful that God woke me up one morning and put my feet in a different direction, leading me into a new career. I cannot wait to see what you do next, Lord.

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ABSTRACT

Cystic fibrosis (CF) is a heritable monogenic condition with allelic heterogeneity. A variety of sequence alterations in the cystic fibrosis transmembrane conductance regulator (*CFTR*) gene results clinically in multisystem disease including a reduced reproductive capacity in affected males. Knowledge of reproductive capacity is often disclosed to parents by medical providers and to affected male children by medical providers and their parents. Despite advanced technology and therapeutics yielding the benefits of increased life expectancy and quality of life, males affected with CF largely remain uninformed in broader areas of sexual health and particularly in how their concept of masculine identity can be negotiated within the context of this inherited syndrome. Using a descriptive retrospective quantitative approach, this preliminary study examines and reflects on sexual education provided to males with CF by their fathers during their adolescence. The purpose of the study was to establish themes from interviewing affected adult males and/or fathers with an affected male child to begin the development of standards promoting more effective genetic counseling communication practices for an often-ignored population. The purpose was also to lay a foundation for generating resources for males with CF regarding their reproductive and sexual health (RSH).

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LIST OF ABBREVIATIONS

CAVD	Congenital Absence of the Vas Deferens
CF.....	Cystic fibrosis
CFF	Cystic Fibrosis Foundation
<i>CFTR</i>	Human Cystic Fibrosis Transmembrane Conductance Regulator gene
CFTR	Human Cystic Fibrosis Transmembrane Conductance Regulator protein
GC.....	Genetic Counselor
HCP.....	Health Care Professional Provider
ICSI.....	Intracytoplasmic Sperm Injection
IRT	Immunoreactive trypsinogen
IVF	In vitro fertilization
ODPHP	Office of Disease Prevention and Health Promotion
PI.....	Principle Investigator
PTSD.....	Post-Traumatic Stress Disorder
RSH.....	Reproductive and Sexual Health
STI	Sexually Transmittable Infection
US	United States

CHAPTER 1: BACKGROUND AND LITERATURE REVIEW

1.1 A CYSTIC FIBROSIS (CF) PRIMER

1.1.1 Defining and Identifying the Need for Sex Educators for Males with CF

The United States (US) Office of Disease Prevention and Health Promotion (ODPHP) Healthy People 2020 agenda has eloquently set forth a national edict of improving reproductive and sexual health (ODPHP, 2018). Healthy People 2020 focuses on reproductive and sexual health (RSH) as crucial to eliminating health disparities, reducing rates of infectious diseases and infertility, and increasing educational attainment, career opportunities, and financial stability (ODPHP, 2018). Improving individual reproductive health builds on the foundation of what has been established physically, biologically, and socially in childhood and adolescence. The process of understanding personal RSH is a defining developmental milestone in adolescence and represents a series of complex biopsychosocial behaviors. While questions of what to include in conversations about sexual development, and when and how this information is to be disseminated is not easily answered, the responsibility of who should have the discussion is clear: parents. Parents are the choice of children and experts alike (Flores & Barroso, 2017; Breuner, et al., 2016; Wyckoff, et al., 2008).

Parents are traditionally the first point of contact for sexual education and therefore have a principal role to play in educating their children on RSH regardless of whether a genetic condition is present or not (Breuner, et al., 2016; Wyckoff, et al., 2008). Parent-child communication about sex, at its best, is bidirectional communication

between parents (or a parental figure) and the children about any issues regarding reproduction, sexuality, and related outcomes, including dating and emotional intimacy. Parents have been identified as the ideal sex educators for their children because they can reach youth early and can provide sequential and time-sensitive information responsive to an adolescent's questions and anticipated needs (Krauss & Miller, 2012). The RSH of most adolescents is greatly influenced by the powerful role of parents in their children's sexual socialization because parents are influential in shaping adolescent sexual decision-making (Diiorio, Pluhar, & Belcher, 2003).

Wyckoff and colleagues (2008) discovered that most parents indicated a willingness to become sex educators for their children but also have some reservations. Despite parents' willingness to function as sex educators covering wide topics of RSH, parents have also described apprehension about initiating a discussion without the child's prompting (Baier, Margaret, & Wampler, 2008); emphasizing sex only in terms of consequences and cautionary statements (Jerman & Constantine, 2010); and children viewing any communication about sex as a green light to become sexually active (Meshke & Dettmer, 2012). Both fathers and mothers interpret sexual education as giving "the talk" focused on basic reproduction rather than an informal, ongoing, and comprehensive approach to sexual education that includes love, dating, gender roles, and body image. Both fathers and mothers have expressed a desire to provide information to their children regarding the relational aspects as well as the biological process of sexuality. However, some parents reported embarrassment and lack of necessary sexual health knowledge, rendering them inefficient to have the conversations they wanted around sensitive and uncomfortable topics (Ballard & Gross, 2009).

How does sexual education differ for a child with a genetic condition? For families impacted by genetic conditions that include infertility, there are added concerns for RSH discussions that may include the emotional, financial, and social costs that accompany having the genetic condition and having these discussions with a potential partner. Conversations may include discussing associated genetic disease morbidity and associated reduced life expectancy. Children with genetic conditions, like other children, must learn to balance the benefit of social connection versus the risk of rejection as they negotiate dating and peer relationships. The questions of who contributes to their knowledge, what is said, and what are the personal and public health impacts remain largely unanswered. In an age of genomic medicine and precision healthcare, these questions prompt investigation. Furthermore, identifying auxiliary providers who can further educate parents and can facilitate sexual education is equally salient.

One example of a Mendelian genetic disorder where the aforementioned considerations intersect is cystic fibrosis (CF). CF is a complex multi-system disorder typically characterized by progressive pulmonary disease, nutritional disease due to pancreatic insufficiency and male infertility due to congenital absence of the vas deferens (CAVD) (Online Mendelian Inheritance in Man [OMIM] 219700, 2019). CF is one of the most common genetic disorders, yet knowledge of a child's diagnosis does not automatically confer parents or family with the proficiency to discuss complex concepts of RSH. Adolescent males with CF are a unique group for several reasons:

1. Parents and /or health care providers may make males with CF aware of their infertility during their adolescence.

2. Males with CF are likely to develop romantic relationships with the knowledge of or the expectation of CF related infertility before entering a romantic relationship (in contrast to men who may discover their infertility while in a relationship).
3. Males with CF are living with syndromic, multi-system, life-limiting disease for which infertility is just one of many complicating signs of the condition.

1.1.2 Cystic Fibrosis Background and Pathophysiology

Cystic fibrosis (CF) is one of the first genetic conditions identified and described in the literature. Extensive research, policy, publication, capital, public health, and precision medicine attention has been devoted to the natural history and progression of CF, the clinical pathophysiology and disease management, the molecular genetics mechanisms, the economic burdens, its therapeutics and a plethora of other challenges associated with the disease. CF and CF-related CAVD are multisystem diseases with ill-defined genotype-phenotype relationships (Drumm, Ziady, & Davis, 2012; Paranjape & Zeitlin, 2008). Infertility stems from a blockage, atrophy, or an absence of the vas deferens, the structures responsible for delivering sperm from the testis into the semen. Hence, during intercourse, fertilization is not achieved without adequate sperm concentration in the seminal fluid (Sharma, 2019; Ong, et al., 2017; Katkin, 2012; Hayes & Savage, 2008).

1.1.3 Genetic Underpinnings, Etiology and Mutational Class

CF is a heritable autosomal recessive monogenic disorder caused by mutations in the cystic fibrosis transmembrane conductance regulator gene (HUGO Gene Nomenclature Committee approved gene symbol, *CFTR*) with *cytogenetic location*:

7q31.2 and genomic coordinates (from GRCh38): 7:117,480,024-117,668,664 (OMIM 219700, 2019; OMIM 602421, 2019).

In CF and CF associated CAVD, more than 2,000 reported sequence alterations in *CFTR* yield a robust pathogenic mutational and phenotypic spectrum (OMIM 602421, 2019; Cystic Fibrosis Mutation Database, 2011). Pathogenic *CFTR* alleles impact the anatomy, production, location processing, and/or functioning of *CFTR*. However, in the presence of, and even in the absence of CF, it has been written that most males with *CFTR* variants are infertile regardless of the disease severity and genotype (Drumm, Ziady & Davis, 2012).

1.1.4 Epidemiology of Cystic Fibrosis

Cystic fibrosis is a relatively common genetic condition affecting ~1 in 2500 individuals of European descent. There are approximately 30,000 people in the United States living with CF and about 70,000 people living with CF worldwide. (Cystic Fibrosis Foundation [CFF] Patient Registry, 2018; Katkin, 2012; O’Sullivan & Freedman, 2009).

1.1.5 Diagnosis and Testing

Carrier testing is available for at-risk individuals, and targeted molecular analysis is available when a shared/familial variant is known (Ong, et al., 2017). Also, all fifty states and the District of Columbia’s newborn screening programs perform immunoreactive trypsinogen (IRT) testing for CF (Ross, 2008). The median age at diagnosis of CF is six to eight months and over 75% of people with CF are diagnosed by two years of age (Sharma, 2019). A well-performed and interpreted chloride sweat test is the gold standard for accurate diagnosis of CF, but the disease can also be screened for

and diagnosed by a combination of genetic analysis, biochemical assessment, and clinical presentation (Ong, et al., 2017; Dequeker, et al., 2009).

1.1.6 Prognosis and Therapy

The median predicted survival for CF patients born in the US in 2018 is 47.4 years (95% CI, 44.2-50.3) and the median survival age is higher in males than in females. (CFF Patient Registry, 2018; Katkin, 2012; O’Sullivan & Freedman, 2009). In addition to CFTR modulator therapies, symptomatic care is supportive. Documented best practice guidelines, such as assisted reproductive technologies, are employed for male infertility.

1.2 FAMILY CONSIDERATIONS FOR A CHILD WITH CF

Health beliefs are but one traditional foundation that families maximize and preserve identity, maintain connectedness, seek informed care, and choose and respond to testing for and managing illness. The meaning of the illness experience, beliefs of what is possible, and the roles a parent takes will vary with cultural norms and other socioeconomic determinants such as access, exposure, and intent. Some cultures tend to be more fatalistic than others, while some stress personal responsibility and agency (Rolland, 2006). Maintaining vigilance for signs of CF-related acute illness is associated with high treatment and emotional costs (Barker & Quittner, 2016; Hassan, et al., 2018; Hayes & Savage, 2008; Nagy & Ungerer, 1990; Ouyang, et al., 2009; Quittner, et al., 2016; Shardonofsky, et al., 2019). Diagnostic results and the anticipation of long-term health consequences can disrupt core family beliefs and can create a fundamental betrayal of trust in how parents perceive their bodies and or the classically widely held perception of pediatric invulnerability (Kleinman, 1988). Bellato and colleagues (2016) report that families caring for a child with a chronic condition are vulnerable, after the initial

diagnosis, and remain vulnerable throughout that child's lifetime. This vulnerability can be related to a lack of support from health service professionals.

As will be covered separately below, the diagnosis of chronic illness or a genetic syndrome in a child has a specific and unique emotional impact on fathers as they ponder and plan for expectant care routines of administering medications, performing intensive chest physiotherapy, and monitoring the pulmonary and the nutritional status of CF (Nagy & Ungerer, 1990). As a parent, fathers will also likely be confronted with the average annual cost of care for a person with CF in the US being over 20 times higher than caring for someone without CF (Ouyang, et al., 2009). Health care resource utilization is high with approximately 47% of the population with CF requiring at least an annual inpatient admission, and an additional 27% having subsequent hospitalizations, averaging 29 days per year in a hospital (Hassan, et al., 2018).

1.3 THE EMERGING ROLE AND VISIBILITY OF FATHERS

1.3.1 The Impact of Fathers' Perceptions

Men and women have different perceptions of the significance of a genetic condition and consequent care of a family member and family unit that are impacted by a genetic condition. Although the fields of public health, clinical medicine, and genetic sciences are shifting, current social practice assigns women disproportionate responsibility for family care in many regards. Family care, however, should not be solely a woman's responsibility (D'Agincourt-Canning, 2001). Fathers show evolution in adapting to evolving best evidence approaches to various aspects of comprehensive CF care and, as such, are critical in family planning and family health (Herrick, et al., 2004; Swallow, et al., 2012). As family systems in the US change via social, political,

technological and economic factors, researchers are increasingly exploring how fathers combine their physical and emotional presence with their experiences of economic and social responsibility (D'Agincourt-Canning, 2001; Dyson, Berghs & Atkin, 2016; Herrick, et al., 2004; Kobylanskii, et al., 2018; Kotelchuck & Lu, 2017; Shardonofsky, et al., 2019; Wolff, et al., 2010). Sensitivity to a father's experience in his child's healthcare, from preconception care to pediatric involvement, can support his psychological maturation and the various roles he can play in the overall family structure (Kotelchuck & Lu, 2017).

Fathers should be encouraged to be conscious and positive participants at patient appointments. Some fathers have reported that the medical system can be alienating in a pediatric and prenatal setting, and it can hinder their involvement, diminish their perceived significance and shrink the importance of their contribution (Atkin, Berghs, & Dyson, 2015; Dyson, et al., 2016; Hovey, 2003; Nagy & Ungerer, 1990; Pelentsov, Laws, & Esterman, 2015; Priddis, et al., 2010; Wolff, et al., 2010; Wolff, et al., 2011). For some fathers, more explicit descriptions of their children's genomic information may help promote a sense of competency and mastery in the home and in the care of their child (D'Agincourt-Canning, 2001; Herrick, et al., 2004; Hovey, 2003; Rolland, 2006). Fathers desire, and are open to communication from health care professionals (HCPs) and from other fathers, but may ask fewer questions (Kobylanskii, et al., 2018; Pelentsov, et al., 2015; Wolff, et al., 2011). In cases of genetic conditions and chronic illness, fathers have been observed, if needed, to produce care that extends past normative structures of support (Santos, et al., 2018). Of special significance, considering that last point is that

now more than any other time in history, more families are being headed by single fathers (Wolff, et al., 2011).

1.3.2 The Emotional Impact of Genetic Conditions in Children on Their Fathers

Rivard & Mastel-Smith (2014) reported four examples of the specific and unique emotional ways fathers can be impacted by a child with a genetic condition: 1) fathers' reactions to childbirth and disability are different from those of other family members; 2) paternal stressors center on financial issues, family commitments, and feelings of incompetence with the child who has a genetic condition; 3) fathers' need for education about causes of genetic disease, care of the child, and prognosis are not adequately being addressed; and 4) paternal responses, including stress and guilt, have a great impact on the child with a genetic condition. Several studies have highlighted these same concerns specific to CF. For example, several researchers reported that fathers articulated feeling overwhelmed and isolated, yet hopeful, as they draw strength from their child (Hayes & Savage, 2008; Priddis, et al., 2010; Shardonofsky, et al., 2019). Irish fathers expressed constant daily worry about immediate threats to their child's health, which kept them "on their guard" (Hayes & Savage, 2008). The Hayes and Savage (2008) research also found that some fathers indicated an expectation that "they must be stronger" and one father shared that he is never asked how he manages: "I mean I do not want sympathy or anything like that but they (people) just don't (ask)" (Hayes & Savage, 2008).

When speaking about affected children, fathers' intense emotions and reflective body language are documented repeatedly. Fathers who take care of children with genetic disorders worry and are concerned (Bellato, et al., 2018; Hovey, 2003) with sometimes lower acceptance of their child than fathers living in an inner-city (Kobylianskii et al.,

2018). When the emotional valance is high, the pendulum can swing further, resulting in grief, depression, or PTSD, however, if channeled differently, strong emotions in fathers can promote an emerging strength of advocacy for their child. In several studies of fathers with children affected with a genetic illness, fathers wanted more information about the genetic condition and genetic risk information from HCP than they received. They requested foundational knowledge about their child's expected challenges and losses associated with their genetic condition (Atkin, et al., 2015; Dyson, et al., 2016; Hovey, 2003; Pelentsov, et al., 2015; Rivard & Mastel-Smith, 2014). Fathers who experienced a shock after a child's diagnosis needed anticipatory guidance. However, multiple factors require interrogation and integration. Unfortunately, often fathers described feeling "invisible" to HCPs, sharing concerns of perceived gender-biased encounters and environments (Atkin, et al., 2015; Dyson, et al., 2016; Kotelchuck & Lu, 2017; Rivard & Mastel-Smith, 2014). Through a meta-analysis of parental needs, fathers expect inclusion in the process of genetic counseling, engagement in information sessions and support groups, and clear instruction on treatment protocols for their children. They also expect anticipatory guidance across transitional periods of their children's growth and development from all HCPs (Pelentsov, et al., 2015).

What can be surmised from the literature is that paternal support does not diminish the uncertainty of disease variability and prognosis, but it facilitates family adaptation. As a father is navigating the unknowns of a chronic illness with his child, he can equip his son to navigate uncertainties inherent in genetic information and disclosure. Fathers can have a positive impact by building their child's self-concept and identifying how best to combat social stigma and self-consciousness associated with genetic

conditions (Derlega, et al., 2014). Fathers may even improve decision making regarding reproductive options and compliance with treatment, especially where non-compliance is present or a significant concern.

1.4 THE CURRENT STATE OF FATHER-SPECIFIC SEXUAL EDUCATION

Santos and colleagues' (2018), research demonstrated that fathers as caregivers operate across affective, physical, social, and relational domains. An additional domain often navigated, is sexual education, whether or not a child has a known genetic condition (Coakley, et al., 2017; e Silva, et al., 2016; Murphy, et al., 2016) because humans are sexual beings irrespective of other challenges (Murphy, et al., 2016). In fact, there has been a steady increase in the research investigating RSH "able-bodiedness" with children who have genetic conditions, but there is little research exploring fathers' comfort levels, motivation, or informational needs surrounding sex education for their children (Flores & Barroso, 2017).

However, while extant literature on parent-child communication is present, there is less information on father-child sexual education communication. Yet fathers' perspectives are vital to understand the range of family functioning for a child with a genetic condition and provide a more robust assessment of communication in a family with a child who has a genetic condition (Wolff, et al., 2010). Most sexual communication studies still document normative sex discussions performed along gender lines in common dyads of father to son and mother to daughter (Wilson & Koo, 2010). Although some studies have reported that male children obtain information from both parents (Wilson & Koo, 2010), there is other literature to support that the depth of

conversation or number of topics discussed is higher among same-sex dyads (e Silva, et al., 2016; Kapungu, et al., 2010; Tobey, et al., 2011).

In the case of CF where there is known infertility or frank sterility, it is important to know what paternal messages are being conveyed by fathers who are tasked with counseling a child on something he, himself, may not have experienced. Some variables may influence how a father guides or fails to navigate his son including a father's early experience of his child's diagnosis, his belief maladaptation to diagnosis, his belief that caregiving is primarily the mother's responsibility, his lack of clarity on his role, and his unmet informational needs. There is a paucity of studies investigating the impact of the role of father-son communication regarding RSH in CF.

1.5 REPRODUCTIVE AND SEXUAL HEALTH PERCEPTIONS MALES WITH CF

In the 1980s, few CF patients lived beyond their second decade of life. But today, life expectancy is approximately 47.4 years (CFF, 2018). Besides, recent family planning technology has made having biological children possible for males with CF who are infertile. Therefore, males with CF can expect to have biological children notwithstanding their genetically dictated reduced reproductive capacity. These expectations are further supported by Thickett and colleagues (2001), who reported that 68% of males with CF expressed a desire to have children and that 43% report that they would consider fertility treatments. Fair and colleagues (2000), reported having children was important to 85% of men with CF. But there are gaps in the timing of when sexual education is disseminated.

Additionally, males with CF demonstrate gaps in their knowledge and misconceptions about RSH in CF. An early study of HCPs from four CF clinics revealed that only 38% reported speaking about using condoms with their CF patients (Sawyer, Tulley, & Collin, 2001). Sawyer and colleagues (2001) documented that only 19% of HCPs reported discussing reproductive options and 13% reported offering semen analysis. By contrast, a later survey of RSH in males with CF reported the mean age when they were informed of their infertility was 16.4 years, yet they also reported that they wanted to have this discussion at 14.4 years of age (Sawyer, et al., 2005). In this study, the average age of sexual intercourse for the males with CF was 17.9 years of age, and two-thirds of them reported that they wanted more information on reproductive options than what they received. This is congruent with results from another study, where 74% of males with CF stated 17 to be the most appropriate age to be offered semen analysis (Rodgers, Baldwin, & Knox, 2000). Frayman, et al., (2008) reported that 19% of parents of sons with CF reported subsequent conversations with a healthcare provider after their son's infertility was disclosed. Parents reported that they initiated this conversation and expressed a desire for more ongoing conversations with HCPs regarding sexual health.

At the time this manuscript is being written, a semen analysis is a much more common practice for CF males than at the time when those early studies published data on how often providers offered such testing, but today, gains in standards of care offering testing are ahead of standards of practice in this area. For example, there are now commercially available at-home sperm analysis kits and there are published recommendations to specialists for semen analysis (Fainberg & Kashanian, 2019; Farber,

et al., 2019). A review of the literature reveals that sex educators and resources for males with CF are not current, not readily available and/or are plagued by misconceptions and confusion.

There are additional misconceptions reported in the literature: 33% of the men assumed they did not need to use a condom, and surprisingly, those informed about sexuality by HCPs were more likely to assume this than those educated by their parents; 10% of the men confused infertility with impotence (Sawyer, et al., 2005; Sawyer, et al., 2009); patients overestimated the risk of their biological child having CF after a successful conception using assisted reproductive techniques and the majority of HCP routinely informed adolescent boys that their sexual performance would not be affected by CF, but only 3% were informed about hypospermia and low (less than 1.5mL) ejaculate volume (Popli, Bourke, & Stewart, 2009).

What we can ascertain from the misconceptions is that while men seem aware and educated about the existence and cause of their infertility, they are under-informed about other biopsychosocial aspects of their RSH. Furthermore, studies have examined RSH, but as of now, little is known about how CF patients are informed and educated about their RSH from childhood into adulthood (Haverman, et al., 2011). As the quality of life increases for individuals with CF, the conversations with children regarding RSH needs to be expanded between HCP, parents, and male children with CF to close the gap between RSH information given and the evidenced-based RSH that satisfies or exceeds ODPHP standard to improve RSH for all.

1.6 GENETIC COUNSELOR – CLIENT COMMUNICATION

In the most recent definition of genetic counseling from the National Society of Genetic Counselors' (GC) Task Force report, genetic counseling was defined as the process of helping people understand and adapt to the medical, psychological and familial implications of genetic contributions to disease (Resta, et al., 2006). GCs can help frame realistic messages to help families and specifically fathers gain a sense of coherence by recasting a medical crisis as a comprehensible, manageable, and meaningful challenge to tackle.

In an era when people are living longer and more time is being spent living with chronic illnesses, understanding genetic risk factors is more valuable to families now than in the past. GCs can assist families by clarifying the nature of CF and by explaining expected challenges that will be faced throughout the condition particularly as it relates to navigating CF-related infertility biology and appraising treatment options with patients. GCs also recognize and consider culture, values, family structure, and other social/relational ideologies necessary for effective communication with patients and families. Professional genetic counseling standards connected with unique positions from other disciplines such as the theoretical learning sciences and public health can inform genetic counselors as they seek to help fathers provide specific sexual education to their son with CF.

Anticipatory guidance regarding childhood development and reproductive information for CF parents is essential. Children who live with infertility-related phenotypes or who have sexual concerns specific to their genetic illnesses rely on parents or health professionals to educate them about their options, yet parents can feel ill-

equipped and overwhelmed (Ballard & Gross, 2009). Creating a scientific, realistic, and empowering narrative for a child with a genetic disorder poses a challenge for parents. GCs are trained to provide anticipatory guidance in addition to educating patients about testing options, diagnosis, and risk assessments. GCs are trained to sensitively employ integrated approaches with cognitive and affective styles to reduce the complexity of communication and balance the delivery of unhurried, clear, and comprehensive messages. GCs and family members can be natural allies on a child's healthcare team, including implications for RSH, as the child's clinical course evolves.

1.7 RATIONALE

There is a surge of awareness of men's pre-diagnostic and post-diagnostic conception of their health where dyadic intergenerational studies demonstrated that health does not only flow from men to their children but also from children to their fathers (Kotelchuck & Lu, 2017). And encouragingly, there is a recent and ongoing conversation on RSH issues in the U.S. in the context of parent-child interactions when CF is diagnosed. Males with CF are living longer and are enjoying a higher quality of life and they are better informed than in the past about their genetic diagnosis and as a result, they are asking more questions about recently developed reproductive options and their RSH (Yoon, et al., 2019).

Despite this, when studied, men continue to report confusion regarding when their reproductive capacity or infertility status was or is to be disclosed, they report misconceptions about condom use and the need for preventing sexually transmittable infections (STI), and they have unanswered questions about how CF impacts their self-identity and the biological norms of sexual functioning (Yoon, et al., 2019). These

misconceptions impact disease incidence and prevalence data as well as the quality of life for men with CF. This study aims to further illuminate the misconceptions and the unanswered questions that persist. And because parents are often the earliest purveyors of the specific RSH information for adolescent males with CF, it seems prudent that parents must be equipped with evidence-based data to successfully pilot this role. This study also seeks to categorize the components of a comprehensive discussion of RSH for a male with CF and purposes to establish preliminary data that can be used to refine a construct of a genetic counseling intervention to support father/son with CF when communicating RSH.

1.8 STUDY AIMS

Examining and contributing to these nascent RSH conversations is an attempt to refine the current problems and to guide intervention offered in transitional genetics clinics. Examining father-son RSH can significantly help fathers understand and adapt to medical, psychological, and familial implications of genetic contributions to disease as they prepare to raise their sons with CF in the United States.

As such, the proposed research attempts to address unmet needs in scholarly research and in clinical practice. The goal of this research is to analyze center-specific data to inform genetic counseling, care planning conversations, and shared decision-making about RSH conversations for males with CF. In summary, the aims of this project are threefold:

1. Elucidate the nature of the RSH information desired and needed by males with CF;

2. Catalog what RSH elements are deemed relevant to father-son RSH communication at the patient level;
3. Expand knowledge and understanding of genetic counseling for males with CF in transitional care periods of adolescence and adulthood.

CHAPTER 2: RESEARCH MANUSCRIPT

2.1 INTRODUCTION

The United States (US) Office of Disease Prevention and Health Promotion (ODPHP) Healthy People 2020 agenda has eloquently set forth a national edict of improving reproductive and sexual health (ODPHP, 2018). Healthy People 2020 focuses on reproductive and sexual health (RSH) as crucial to ending health disparities, reducing rates of infectious diseases and infertility, and increasing educational attainment, career opportunities, and financial stability (ODPHP, 2018). Improving individual reproductive health builds on the foundation of what has been established physically, biologically, and socially in childhood and adolescence. In fact, the process of understanding personal RSH is a defining developmental milestone in adolescence and represents a series of complex biopsychosocial behaviors. While questions of what to include in conversations about sexual development, and when and how this information is to be disseminated is not easily answered. However, the responsibility of who should have the discussion is clear: parents. Parents are the choice of children and experts alike (Flores & Barroso, 2017; Breuner, et al., 2016; Wyckoff, et al., 2008).

Parents are traditionally the first point of contact for sexual education and therefore have a principal role to play in educating their children on RSH regardless of whether a genetic condition is present or not (Breuner, et al., 2016; Wyckoff, et al., 2008). Parent-child communication about sex, at its best, is bidirectional communication

between parents (or a parental figure) and the child about any issues regarding reproduction, sexuality, and related outcomes, including dating and emotional intimacy.

But how does sexual education differ for a child with a genetic condition? Men and women have different perceptions of the significance of a genetic condition and consequent care of a family member and family unit that are impacted by a genetic condition. As family systems in the US change via social, political, technological, and economic factors, researchers are increasingly exploring how fathers combine their physical and emotional presence with their experiences of economic and social responsibility. Sensitivity to a father's experience in his child's healthcare, from preconception care to pediatric involvement, can support his psychological maturation and the various roles he can play in the overall family structure (Kotelchuck & Lu, 2017).

However, while extant literature on parent-child communication is present, there is less information on father-child sexual education communication. Most sexual communication studies still document normative sex discussions performed along gender lines in common dyads of father to son and mother to daughter (Wilson & Koo, 2010). Although some studies have reported that male children obtain information from both parents (Wilson & Koo, 2010), there is other literature to support that the depth of conversation or number of topics discussed is higher among same-sex dyads (e Silva, et al., 2016; Kapungu, et al., 2010; Tobey, et al., 2011).

One example of a Mendelian based genetic disorder where RSH considerations intersect is cystic fibrosis (CF). CF including CF with congenital absence of the vas deferens (CAVD) is a disorder with increased morbidity and reduced life expectancy (Online Mendelian Inheritance in Man [OMIM] 219700, 2019). Diagnosis of CF disease

or carrier risk status does not automatically confer parents or families with the proficiency to discuss complex concepts of RSH. Individuals with nonlethal CF phenotypes reach an age of reproductive fitness but males with CF possess up to a 98% reduced or absent reproductive capacity posing some unique challenges such as:

1. Parents and /or health care providers may make males with CF aware of their infertility during their adolescence.
2. Males with CF are likely to develop romantic relationships with the knowledge of, or the expectation of CF-related infertility before entering a romantic relationship (in contrast to men who may discover their infertility while in a relationship).
3. Males with CF are living with syndromic, multi-system, life-limiting disease for which infertility is just one of many complicating signs of the condition?

How does sexual education differ for a child with a genetic condition? For families impacted by genetic conditions with a nonlethal phenotype, but includes infertility, there are added concerns for RSH discussions that may include the emotional, financial, and social costs that accompany having the genetic condition, and having these discussions with a potential partner. Conversations may include discussing associated genetic disease morbidity and associated reduced life expectancy. Children with genetic conditions, like other children, must learn to balance the benefit of social connection versus the risk of rejection as they negotiate dating and peer relationships. The questions of who contributes to their knowledge, what is being said, and what is the personal and public health impact are unknown but requires study in an age of genomic medicine and precision healthcare. Furthermore, identifying auxiliary providers who can further educate parents and can facilitate sexual education equally is salient.

In the case of CF where there is known infertility or frank sterility, it is important to know what paternal messages are being conveyed by fathers who are tasked with counseling a child on something he may not have experienced. Some variables may influence how a father guides or fails to navigate his son including a father's early experience of his child's diagnosis, his belief maladaptation to diagnosis, his belief that caregiving is primarily the mother's responsibility, his lack of clarity on his role, and his unmet informational needs. There is a paucity of studies investigating the impact of the role of father-son communication regarding RSH in CF.

In summary, this thesis will explore what impact, if any, CF has on father-son communication regarding RSH. The goal of this research is to analyze center-specific data to inform genetic counseling, care planning conversations, and shared decision-making about RSH conversations for males with CF. Its aims are threefold:

1. Elucidate the nature of the RSH information desired and needed by males with CF;
2. Catalog what RSH elements are deemed relevant to father-son RSH communication at the patient level;
3. Expand knowledge and understanding of genetic counseling for males with CF in transitional care periods of adolescence and adulthood.

2.2 METHODS

2.2.1 Theoretical Approach

This preliminary research is a descriptive quantitative study that involved coding of responses to an online questionnaire.

2.2.2 Participant Recruitment

Participants were recruited from the University of North Carolina Chapel Hill Cystic Fibrosis Center (UNC) and through social media support groups. Recruitment efforts included flyers, referrals from providers at clinics, and word of mouth. The men at UNC were invited to participate in an online questionnaire. Adult males who had posted CF content over the past six months across six Facebook support groups (a family support group, a parent support group, a rare mutation group, a late diagnosis group, and two general CF groups) were contacted to participate. After obtaining host permissions, invitations were made to 50 men via Facebook Messenger, and 25 clinic participants were provided with program flyers directly through clinic staff.

Eligibility criteria included males aged 18 years or older with a documented personal history of CF. Participants were required to identify as speaking and reading English fluently. Participants were excluded if they did not meet these inclusion criteria, if they had previously had genetic counseling by this study's principal investigator (PI), or if they were unable to give informed consent. All surveys were completed between January 2020 and April 2020.

2.2.3 Data Collection

Questionnaires were constructed and offered using the Qualtrics^{XM} survey software that provided completion online using a computer or a mobile device.

The instrument for adult males included eight genetic conditions questions, five questions on the diagnosis of CF, 22 questions on sexual education, 15 questions on RSH communication, and seven questions on demographics (See Appendix A). After communicating the study risks and benefits, participants were directed to open the link to

the questionnaire and by answering a single question, participants completed the informed consent process. Upon completion of the questionnaire, participants were given the option to provide their contact information for follow up communication to clarify questions if needed. Participants received a Visa gift card in the amount of US \$20.00.

2.2.4 Data Analysis

Thematic analysis methods were used for identifying consistent ideas across the participants' responses. Attempting coding and thematic analysis was utilized to frame the results of the questionnaires because it is a flexible method allowing for a variety of viewpoints (Braun & Clarke, 2006) and can be used to identify patterns in the data. There were no preset themes for the study's focus. Summary statistics (frequencies and percentages) of the emerging categories and themes were computed.

2.3 RESULTS

2.3.1 Response Rate

The average time spent on each questionnaire was 33 minutes. Men in this study were initially asked to complete an in-person interview, but there were no respondents after six months despite marketing through flyers and point persons at seven local community resources. In the revision of the delivery model, an online questionnaire was constructed and refined by professional staff specializing in the areas of Marriage and Family Therapy, Developmental Psychology, Research Methods, and Cystic Fibrosis/Pulmonology Genetics. The questionnaire was devised with both male and female input across these specialties.

Cystic Fibrosis Center Staff and the primary investigator were asked to personally introduce the questionnaire to men who engaged in services through their CF clinic to

obtain a sample of adult males with CF or father of sons with CF. Twenty-two male participants and seven fathers responded to the questionnaire link through online and site-specific research study promotion. Six adult male participants and four father participants were considered ineligible due to an item non-response rate of 80% or above. Since only two fathers completed the questionnaire it was determined that the data for fathers was insufficient to provide an adequate comparison group. Thus the final analytic sample consisted of 16 men with CF.

2.3.2 Demographic Data

Twelve (75%) of the participants identified as White/Caucasian, one participant (6%) identified as African American, one participant (6%) identified as Hispanic/Latino, one participant (6%) identified as American Indian/Alaskan Native, and one (6%) participant identified as biracial. Seven (44%) participants had obtained a four-year-degree, and six (38%) of the participants reported some college. One participant (6%) reported a doctoral degree and one participant (6%) reported a professional/advanced degree. Half of the participants (50%) participants reported they were married. Five (31%) participants reported that they were in a committed relationship, and three (19%) participants reported being currently single. Mutational differences and other demographic data are presented in Table 2.1. Where impactful, mutation data is separated from overall averages to enrich participant nuances. Also, two participants reported that they did not grow up with their fathers.

Table 2.1 Demographics Chart of CF Adult Males

Demographics	N	%
Age		
18-25	3	19
26-33	6	38
34-40	4	25
41-47	1	6
48-54	0	0
54-61	1	6
62-67	1	6
Ethnicity		
Black	1	6.25
Bi-Racial	1	6.25
Native American	1	6.25
Latin American	1	6.25
White	12	75
Marital Status		
Married	8	50
Committed Relationship	5	31
Single	3	19
Widowed	0	0
Divorced	0	0
Separated	0	0

Table 2.1 Demographics Chart of CF Adult Males

Demographics	N	%
Education		
Less than High School	0	0
High School Graduate	1	6
Some College	6	38
2-year Degree	0	0
4-year Degree	7	44
Professional Degree	1	6
Doctorate	1	6
Genotype		
DF508/DF508	7	44
DF508 + 3120 + 1G>A	1	6
DF508 + G542X	1	6
N1303K/N1303K	1	6
DF508/Unknown Mutation	1	6
Unknown Mutations	2	13
Unlisted Mutations	3	19
Hospitalizations		
0	4	25
1	2	13
2	4	25

Table 2.1 Demographics Chart of CF Adult Males

Demographics	N	%
3	2	13
4	3	19
8	1	6

2.4 QUESTIONNAIRE RESULTS

2.4.1 The Reproductive and Sexual Health Needs of Men with CF

The participants were asked several questions to define their perceived needs as adolescents regarding RSH and what expectations they had of their fathers in educating them. Table 2.2 displays open text responses to the questions of: 1) what participants most wanted to know about RSH and CF as adolescents; 2) what myths participants had as adolescents about CF and RSH; 3) what participants thoughts should be included in RSH communication for young men with CF; and 4) what participants found was most stressful about having CF during adolescence while dating, having relationships and sex. Together these questions elicited 32 topics, or needs, were referenced by the men throughout their responses and are considered significant aspects of CF sexual education for males impacted by this diagnosis. Table 2.3 addressed those perceived topics, and needs, the men felt uniquely belonged in father-son communication for males with CF.

2.4.2 A Sexual Education Program Designed by Men with CF

When asked to design a sexual education program for males with CF, 13 respondents had an overlap of RSH topics, but there were some distinctions among participants. Regarding how best to deliver sexual education messaging, one respondent suggested a video with built-in question and answer sessions as the best strategy. The

remaining 12 respondents reported reproductive, sexual, and psychosocial impact. Four respondents reported that they felt that males should be informed about the low ejaculation volume of men with CF. One respondent relayed the importance of discussing inheritance that a child he has may have CF. Another respondent reported that the logistics and mechanics of infertility needs to be explained. As it related to sexual education, a frank discussion about the impact of CF on a male's sex life should be considered including pros and cons of sex, why safe sex is important for men with CF, the sexual side effects of the CF medications, the importance of pacing sexual activity and lung function, and the "importance of keeping biological agents outside of one's body during sex." Other males included the disclosure of CF status to a partner and exercises to enhance self-esteem and self-confidence.

2.4.3 CF Impact on Dating, Relationships, and Sex

When asked about stressors or benefits because of CF during high school, three participants reported there was no stress for them, stating "my childhood was free of CF restrictions," and I was "treated as a normal student." Others expressed benefits to CF such as, "I could eat anything and stay skinny," and "I could have sex without worries of getting a woman pregnant." However, for ten participants, there were immediate stressors with CF. Four of the participants regretted that they could not play sports, or, if they did, they were "benched" or "missed games." Four participants reported interactions with peers were limited due to "constantly being sick," "all my treatments and vest," having to "be cautious in class" and one participant reported having to "keep CF a secret" during adolescence as a stressor. Other participants cited concerns from parents regarding dating

and parents' perceptions that they would get sick if allowed to date while remaining participants just mentioned absences from school and work.

Table 2.2 Sexual Education Needs During Adolescence Among Males with CF (N=16)

Topic of Interest	DF508/ DF508	DF508 Heterozygotes	N1303K/ N1303K	UNK/ UNL	Total
Early Stressors					
Sick all the time	3	0	1	2	6
Socialization: Friends and Dating	3	1	0	2	6
Finances: Work	1	0	0	0	1
Sports	1	0	0	3	4
Infertility Worry	3	2	0	1	6
“Different”	2	1	1	2	6
No Early Stressors	3	0	1	0	4
Reproductive Health					
Why am I infertile?	2	1	1	4	8
What causes me to be infertile?	2	1	1	4	8
Not everyone is infertile and why?	2	1	1	4	8
How is sperm blocked?	2	1	0	1	3
What is my chance to have a child with CF?	4	0	0	3	7
What is my chance to have a child with CF if I use IVF?	1	0	0	2	3

Table 2.2 Sexual Education Needs During Adolescence Among Males with CF (N=16)

Topic of Interest	DF508/ DF508	DF508 Heterozygotes	N1303K/ N1303K	UNK/ UNL	Total
Sexual Health					
Will sex/breathing the same air as an unaffected woman/taking a shower after sex make me sick?	3	0	0	1	4
How can I make sex meaningful?	0	1	0	1	2
How do I handle sexual abuse?	0	0	0	1	1
What do I do?	0	0	0	1	1
Safe Sex	2	3	0	3	8
How does a woman's bodywork?	0	1	0	3	4
Sexual Performance					
Be honest about changes which can include negative impact (Low ejaculation volume)	2	1	0	1	4
Talk about strategies of pacing, endurance, and stamina	1	1	0	2	4
Address side effects of CF medication to sexual performance	1	0	0	1	2
Medication Maintenance for sexual performance	1	0	0	0	1
How do I approach women?	0	1	0	2	3
How do I treat women?	0	1	0	2	3
How do I disclose my CF status?	2	0	1	3	5
Which Woman will I get?	1	0	0	1	2
Am I dateable?	1	0	0	1	2

Table 2.2 Sexual Education Needs During Adolescence Among Males with CF (N=16)

Topic of Interest	DF508/ DF508	DF508 Heterozygotes	N1303K/ N1303K	UNK/ UNL	Total
Impact of Hospitalizations on Relationships?	0	1	0	1	2
Psychological					
How do I build my esteem with CF/ “unworthy of love”?	1	0	1	1	3
How do I increase my confidence with CF?	0	1	0	1	2

Table 2.3: Father-Son Sexual Education Topics by Mutational Type

Topic of Interest	DF508/ DF508	DF508 Heterozygotes	N1303K/ N1303K	UNK/ UNL	Total
Reproductive Health					
Why am I infertile?	2	1	1	4	8
What causes me to be infertile?	2	1	1	4	8
Not everyone is infertile and why?	2	1	1	4	8
How is sperm blocked?	2	1	0	1	4
What is my chance to have a child with CF?	4	0	0	3	7
What is my chance to have a child with CF if I use IVF?	1	0	0	2	3

Table 2.2 Sexual Education Needs During Adolescence Among Males with CF (N=16)

Topic of Interest	DF508/ DF508	DF508 Heterozygotes	N1303K/ N1303K	UNK/ UNL	Total
Sexual Health					
Will sex/breathing the same air as an unaffected woman/taking a shower after sex make me sick?	3	0	0	1	4
How can I make sex meaningful?	0	1	0	1	2
Safe Sex	2	3	0	3	8
Sexual Performance					
Be honest about changes to my reproductive system which can include negative impacts	2	1	0	1	4
Talk about strategies of pacing, endurance, and stamina.	1	1	0	2	4
Address side effects of CF medication to sexual performance.	2	0	0	1	3
How do I approach women?	0	1	0	2	3
How do I treat women?	0	1	0	2	3
How do I disclose my CF status?	2	0	1	3	6

The mean age of dating was 14 years old. Six participants (38%) reported that CF did not have an impact on their ability to date as adolescents. Four participants (25%) reported “a moderate amount” of impact to dating, two participants (13%) reported “a little” impact on their ability to date, three participants (19%) reported “a great deal” of impact on their ability to date, and one participant (6%) reported “a lot” of impact on their ability to date.

Nine participants (56.25%) reported that CF had a “significant” impact on their ability to have children. Three participants (18%) reported a “moderate” impact on their ability to have children, and three participants (18%) reported “a little” impact on their ability to have children. One participant noted “a lot” of impact on their ability to have children.

Men expressed that despite differences in how CF impacted their dating, there were distinct challenges they encountered psychosocially as they reflected on their experiences. For instance, men reported in adolescence there were restrictions on “dating” and being “sociable.” A respondent reported his hospitalizations significantly impacted dating and that he also felt he did not have adequate time to devote to someone or a relationship. However, some men reported that finding a partner could be difficult because of their intrapersonal feelings. Two respondents reported, “I felt self-conscious and unworthy of love,” and “I wondered if anyone would date (me).” Another expressed concern that people would not understand CF.

Also, men expressed that disclosure was a challenge. One respondent reported he kept his CF and related infertility a secret. Another reported “it’s [sic] hard telling women about this or you get the types who only want you because they think they’re saving you or something,” and another commented “when it came to dating I had to explain my CF before things got serious to make sure they knew what they were getting into. Not a lot of woman can handle that (CF).”

While a majority of the respondents (60%) reported that they did not feel hindered by any sexual myths, participants were uncertain regarding their chances of having a child who would also have CF (N=1, 8%), were unclear as to whether being infertile

meant that a male would produce no sperm (N=1, 8%) and were concerned if “breathing the same air of any girl would make him sick (N=1, 8%).”

When asked what they most wanted to know in adolescence regarding sexual education, there was no majority response. Eleven respondents reported there was a need for CF-specific as well as general sexual education in middle and high school. As it relates to CF-specific knowledge, they reported they wanted to know about their risks for STIs as a male with CF and the impact of CF on the male reproductive system. Regarding general sexual education, they reported wanting more information on how to approach women, how the female body worked, and how to treat a woman the “right way-in a way that would make them feel appreciated.” One respondent reported that his parents were so awkward about sex that it deterred him from wanting to know more.

Three of the 16 respondents simply stated they wanted more information on “everything”, and one of these three men noted that “everything” should be inclusive of topics like rape/sexual abuse of boys that he experienced.

2.4.4 Father-Son Communication: Primary Resources and Primary Sources

How individuals obtained education regarding dating, relationships, and sex varied from person to person. All 16 participants cited multiple sources for their education during the ages of 10-13 years, except for two participants who reported learning about these topics singularly from a sister or another family/community member. Between the ages of 10-13, men reported their primary source of sexual education were friends unfamiliar with CF (N=11, 68%) and personal experience (N=10, 62%). Only 44% of the men reported their father was a primary source of information. Five men (31%) cited that their mothers were a primary source of sexual education and five men

(31%) reported that sexual education classes were their primary source of sexual education. Finally, four men (25%), reported books as their primary source, and three men (19%) selected brothers.

The participants reported that their sources of information for sexual education during the ages of 14-18 years were personal experience (N=12, 75 %), friends unfamiliar with CF (N=7, 43%), and internet websites (N=6, 38%). By ages 14-18, four men (N=25%) reported that their fathers and books were the fourth most-cited sources of information used to learn about dating, sex, and relationships. Other, but less-frequently cited sources of information included brothers (N=3, 19%), sexual education classes (N=3, 19%), and sisters also selected by three men (N=3, 19%).

In summarizing the top three resources the men trusted most for their sexual education as adolescents, 14 men (75%) reported people they dated, seven men (44%) reported friends and six men (38%) reported internet websites. Mothers and fathers were equally cited as fourth in resources they trusted as adolescents. Parents were elected after the internet (N=5, 31%) and parents were preferred over books (N=4, 25%).

Fourteen males reported that their fathers used specific resources to assist them in communication about dating, relationships, and sexual education, sometimes citing multiple resources. Seven men (50%) reported that their fathers used no resource material. Five men (36%) reported the use of personal stories to communicate about dating, relationships, and, sex, and four men (29%) reported faith or religious-based resources. One man (7%) reported the use of a printed book to guide his communications on dating, relationships, and sex.

2.4.5 Topics of Comfort and Discomfort

The participants indicated how comfortable they felt, communicating with their dad, from a range of pre-selected RSH topics. Eleven men answered these questions and marked multiple topics of comfort. Five men (45%) reported that the most comfortable topics to discuss with their father were personal hygiene and how to approach women. Four men (36%) reported that they were able to talk to their dad about being in love, as well as where children come from and how babies were made. Three men (28%) reported being comfortable speaking with their fathers about rejection and self-esteem. Two of eleven men reported feeling comfortable discussing body image (11%), infertility risk (11%), STI/disease prevention (11%), consensual sex/sexual harassment (11%), and handling a breakup (11%). No men voted that it was comfortable speaking with their father about reproductive anatomy, how their bodies worked, masturbation, intimacy, sexual identity, or talking to their doctors about sexual health. Four men reported that they felt comfortable with other topics that were not listed but did not specify or suggest the topic.

Concerning specific RSH topics that the males felt uncomfortable discussing with their father, fourteen males responded to this question. Five men (36%) reported that they felt uncomfortable discussing masturbation. Three men each (21%) reported that they felt uncomfortable discussing with their fathers how their bodies worked, handling a breakup, and different degrees of intimacy in a relationship. In four separate instances, two men (14%) expressed discomfort at mentioning being in love, rejection/self-esteem/belonging, body image, and reproductive anatomy. One participant reported discomfort for each of the following: discussing changes to their body (7%), discussing the “Facts of

Life"/where babies come from, and discussing how to talk to a girl they liked (7%). Five respondents (16%) reported feeling uncomfortable speaking with their dads about masturbation. There was equal discomfort (9%) regarding each of the following topics: discussing breakups, discussing how the body works, discussing STDs, discussing sexual identity, and talking to a doctor about sexual health.

Fourteen respondents provided reasons for why they were uncomfortable with certain topics, that included: the topic experienced was too personal (N=4, 29%) or was described as not the type of things discussed in the home due to "religious reasons" or "not the type of things we talked about explicitly" (N=3, 21%). Four men (29%) reported that their relationships with their father made conversations of this type awkward, (e.g. "my father was never around"). One respondent reported that his father "always wanted me to concentrate on my studies and worried about me falling in with the wrong crowd due to a relationship."

2.4.6 Father-Son Communication: Past Reflections

Of the sixteen participants who responded to the question about their relationship with their father, 50% rated their relationships with their fathers as close and 50% rated their relationships with their fathers as distant. Six men (37.5%) described their relationships with their fathers as "very distant", and two men (12.5%) reported their relationships as "somewhat distant." Conversely, five men (31.25%) described their relationships with their fathers as "very close", and three men (18.75%) reported that their relationships with their fathers were "somewhat close."

Seven men (44%) reported that their fathers could respond to their questions, while nine men (56%) reported that their fathers were not capable of explaining answers to questions that they asked about dating, relationships, and sex.

Fifty percent of respondents reported that their fathers never provided them with helpful information regarding dating, relationships, and sex, 31% reported sometimes, 6% men reported about half of the time, and 12.5% most of the time.

When asked what their fathers said or did that was most helpful in talking about sex, eleven men responded. Five respondents (45%) reported that their fathers kept an open mind and listened to them and their questions. Two respondents (18%) relayed that they were advised on how to approach a girl. One respondent (9%) reported that “he talked to me like a man.” One respondent (9%) reported: “he advised me don’t sleep around make things meaningful.” Six respondents (54%) reported their father did not say or do anything helpful, with one man clarifying, “we talked about anabolic steroids and that’s about it.”

When asked about things said or did by their father that were harmful in their conversations about dating, relationships, and sex, thirteen men responded. Four men (30%) reported that this question was “not applicable.” An additional four men (30%) reported that their father said nothing harmful, and the remaining five men reported limited messaging such as “keep it in your pants.” Two men (15%) cited “lack of communication” as harmful messaging, with one respondent clarifying that “lack of communication forced me to be in the dark and look to other people or classes for answers.” One respondent reported that because his father did not teach him how to deal with his emotions, he acted out impulsively or got himself in trouble when he felt

someone had hurt him, e.g., breaking up with a partner. The remaining respondent reported being told “inappropriate” stories, attitudes, and beliefs about women.

When asked about a time their fathers talked with them about sex, thirteen men responded. Five men (38.4%) reported that they had never talked with their fathers about sex. Two men (15%) reported that they could recall specific situations their fathers spoke with them about sex. Two men (15%) reported their fathers discussed that they need to be responsible and use safe sex, and in the case of one of these individuals, he reported not encouraging his father to expand on the topic any further. Two men (15%) reported limited, unidirectional conversations as a result of their fathers finding them with pornography (e.g. “don’t let me catch you with that (his father’s pornography)” and “he was yelling at me when he found a porno mag in my room and I just listened.”

When identifying what safe sex messages they received, sixteen men responded, ten men reported (63%) “none, safe sex was not discussed.” Of the remaining men who did hear safe sex messaging four men reported they were advised to “wait until...” (25%), and four men (25%) were advised to limit sex partners/practice monogamy. The next most frequently cited safe sex message was to use condoms and to be careful with drugs and alcohol. Additional messaging regarding safe sex was reported by two men who reported that their fathers addressed abstinence as a safe sex practice during adolescence.

2.4.7 Father-Son Communication: Current Reflections

Of the 14 men who responded, four men (29%) reported that their sexual education would have been no different if it was only provided by their fathers. Three men (21.4%) reported that their sexual education would be less complete “I wouldn’t

know much,” “one dimensional narrow in focus,” “less knowledgeable/sex education was more complete.” Three men (21.4%) reported that there would have been no sexual education if left to their fathers alone, with one reporting his father abandoned the family after learning he had CF. Three men (21.4%) reported that their sexual education would have been detrimental if they would have been delivered by their fathers as they described their fathers as “abusive, narcissist” and “heavily into his sexuality.” One respondent reported, “I would have lived a more dangerous sex life.” One man (7%) said he would like to talk with his dad as his dad also has CF and he wants to know how his father met his mother.

Eleven men responded when asked to reflect on how their fathers’ personal beliefs impacted their dating, relationships, and sexual decision making. Four men (36%) reported they do not believe their fathers’ beliefs had an impact on them. One man (32%) reported that he learned from his dad’s mistakes. Two men (18%) listed values and beliefs regarding promiscuity. One respondent reported that if he had listened to his father, he would have slept around more instead of attending church. Another respondent reported his parents were afraid to discuss sex for fear that he would sleep around more.

One respondent reported that he did not know how his father’s beliefs impacted him. One respondent reported his father was absent. One respondent reported that his father emphasized he pursue his education instead of being distracted. One respondent reported that his father’s beliefs and values had a positive impact and that he was able to communicate openly with his dad about love and sex.

2.4.8 Father-Son Communication: Future Reflections

Thirteen men responded with ideas about what should be uniquely included in father-son communication about sex for a child with CF. Four (31%) reported that reproductive information should be included, specifically how sperm was blocked and what reproductive structures are impacted by infertility from CF. Two men (15%) reported that the inheritance pattern both with in vitro fertilization (IVF) and without IVF to determine the risk to a child should be discussed. Two men reported safe sex and meaningfulness of sex in a relationship should be addressed. One reported dealing with women including a need for patience and honesty, not being afraid to show emotion and sexual direction. One male also reported addressing disclosure to a partner. Two men (14%) reported that there needed to be more information on CF and sexual performance. The men expressed concern about performance due to feeling unprepared in this area and wanting more information about topics such as whether bathing before and after sex is necessary, risk of illness from sex, sexual side effects of medication, and endurance, pacing, and stamina required for sexual activity. Their reflections are summarized in Table 2.3.

When asked how their conversations about sex had changed over time with their fathers, twelve men reported (75%) that now that they are no longer adolescents their conversations about sex with their dad have remained the same. One man (6%) reported much better. Two men (13%) reported “moderately better” and one man (6%) reported “slightly better.”

2.4.9 Father-Son Communication in Review

Frequency statistics were used to summarize the men's responses but each piece of the men's sexual communication from open ended and multiple-choice questions was looked at and rebuilt into a holistic narrative to consider overall insights from the individual subject. In Figure 2.1 below, boxes are headed by their question number (Appendix A) at the top in blue and then are fragmented according to the question's sentiment. Applied sentiment scoring is reflected by grey (neutral tone), green (positive tone), orange (negative tone), and yellow (mixed tone) boxes. In the entire dataset, there was a total of 68 negative sentiments coded and there were 31 positive sentiments identified using NVivo12. Each sentiment was reviewed for each case by the PI given NVivo12 is unable to detect sarcasm, double negatives, slang, idioms, and ambiguity.

When NVivo's sentiment analysis was applied to 44 codes (and related subcodes) from 12 questions (For "Q's" see appendix A); Q17, Q19, Q20 (structured), Q20 (open-ended), Q22, Q23, Q24, Q26, Q27, Q31, Q32, and Q34, all about fathers, and father-son RSH communication revealed that more than half, 64% (28/44) of the sentiment codes were negative or very negative (see Figure 2.2). NVivo additionally identified 5 themes and 13 subcodes associated with responses by CF males (Appendix D).



Figure 2.1: Sentiment Analysis of All Coded Male CF Statements. Boxes are headed by their question number at the top in blue and then are fragmented according to the question's sentiment. Applied sentiment scoring is reflected by grey (neutral tone), green (positive tone), orange (negative tone), and yellow (mixed tone) boxes. For example, in the box to the top right, Q12, sentiments had neutral tone, positive tone, negative tone and mixed tone responses.



Figure 2.2: Sentiment Analysis of All Coded Father-Son Communication. Boxes are headed by their question number at the top in blue and then are fragmented according to the question’s sentiment. Applied sentiment scoring is reflected by grey (neutral tone), green (positive tone), orange (negative tone), and yellow (mixed tone) boxes. For example, in the box to the top left, Q20, all associated sentiments had a neutral tone.

2.4.10 RSH Communication: Fertility Knowledge

All males expressed general knowledge of how CF impacted their fertility. One participant reported, he learned “his doctor was wrong” about his infertility status, as he had an unintended teenage pregnancy. The mean age that patients reported that they found out their fertility status was 17, while over 40% of the participants reported that no one told them their status, and/or “they found out on their own.” There was a desire for

disclosure of fertility status by their pulmonologist (N=2, 13%) CF clinic staff (N=2, 13%), and both parents (N=2, 13%). Remaining respondents reported singularly that disclosures were made to them by their brother, mother, and a geneticist.

When asked whose responsibility it is to disclose fertility status, sixteen men responded. Eleven men (69%) reported that it is both parents' responsibility to discuss a male's fertility status if he has been diagnosed with CF. Two respondents suggested a team approach of both the CF team and parents. Three respondents singularly expressed that pulmonologist, genetic counselor, and father should discuss a male's fertility status if he has been diagnosed with CF.

Several themes emerged regarding both helpful and harmful messages received about their infertility. The men described 12 helpful and 11 harmful messages regarding their fertility status with each statement being represented by one participant. The messages most cited as helpful were messages that related their infertility to fathering solutions (46%), such as IVF, testosterone supplementation, adoption, and fostering. The second most cited message was an awareness that safe sex was still important (15%), and the third most cited message was knowing why (the physiological reasons) they were infertile (15%).

There were 12 statements regarding harmful messages received regarding fertility status. Forty-two percent reported that they had not received any messages about their fertility status. Of the 58% that do report harmful messaging, lack of information seemed the most prevalent theme (25%), followed by those who reported they did not physically (physiologically) know why they were not able to father children (17%). Eight percent of respondents reported that their fertility status was withheld as a form of "backup birth

control”, while another 8% identified not being told about safe sex practice as harmful messaging.

2.4.11 RSH Communication and Anticipatory Guidance from Health Care Providers

The men in this study reported a wide range of ages (13-40) for speaking to a CF healthcare team member about their RSH without a parent present, with a modal age of 17 and an average age of 20. Ten of the men reported that their HCPs had discussed sperm analysis with them, and the mean age for this discussion was 24 years old. The ages of respondents who had spoken to their HCPs regarding sperm analysis ranged from 18 to 39 years of age. Eighty percent of those who reported that their doctors discussed this with them said that their providers explained the reasons why sperm analysis was important medical information for them to know.

Regarding speaking with an HCP, 18% reported there was no take-home message that they could identify, 32% reported that they received safe sex messaging about limiting partners and using condoms. Other “take-home” messages included infertility (18%), “be careful but hopeful” (9%), and “typical sexual education message” (9%). Of the respondents who reported there was no take-home message, a respondent disclosed, “We did not address. He asked me if I was sexually active and when I said ‘no’ the conversation moved on.” One respondent reported that his HCP covered sexual education, his fertility status, and inheritance pattern of having a son with CF and he noted that his message had a significant impact on him. “I didn’t want a child to go through what I went through especially with my father who views me as broken and unfit to be his son.”

2.4.12 Family Planning and Family Planning Technologies by Health Care Providers

The men reported a mean age of 25 as the age they began thinking about having a family with a range in age from (13-35). Five men (13%) reported that their HCPs had discussed intracytoplasmic sperm injection (ICSI) with them at a mean age of 21. Eleven men (69%) reported that their healthcare team had never discussed intracytoplasmic sperm injection (ICSI) with them. Men were asked to select who is most responsible to discuss family planning technology. The results were as follows: 25% felt the primary care provider, pulmonologist, genetic counselor, and both parents were equally responsible for discussing family planning technology with men who are diagnosed with CF. While 18.75% of men reported they needed more information before deciding their comfort level, 44% felt comfortable with using family planning technologies like ICSI and 12.5% felt uncomfortable. The remaining 18.75% of participants felt they needed more information before deciding their comfort level. It should be noted that 12.5% were neither comfortable nor uncomfortable with these technologies, and 12.5% just were “never told” anything about family planning technologies like ICSI.

There were several psychosocial considerations listed by the men regarding family planning. Four men were concerned at the “cost of a chance to create life.” While most men were married (50%) or in a committed relationship (31%), they expressed having to have long conversations about risks with their dating partners who were considering families. One man reported “I had the choice to use a medical procedure to collect sperm or use a donor. I just could not bring myself to use mine and then have my kids be a carrier and risk their kids with the same or with CF, too much guilt to do.” The men weighed considerations like their reduced life expectancy when considering family

planning, with one man reporting, “because of the question of life expectancy, I’m not sure if I want to have children with my wife for fear of dying too soon and leaving my wife with the responsibility.” Another stated, “I am not sure I want to do IVF and potentially pass on the CF gene.”

The men were at various stages of resolution with their family planning. One respondent reported that he is homosexual, so his infertility did not impact his family planning. Another male reported he married a woman who had three children and as such reared stepchildren. One respondent reported both he and the woman he married did not want children for several reasons, so it has worked out. Others report that they have no children but “cannot afford to” or that “I have no problem adopting but I would love to have a child of my own.”

2.4.13 Mutational Nuances

In this study, thirteen men responded to the question regarding age at diagnosis. Ten (63%) were diagnosed with CF by the age of 10, and 11 men (85%) by age 13 years. Twelve (92%) were diagnosed by age 19 years. One participant was diagnosed with a DF508 and an unknown mutation at the age of 39.

The mean age of fertility disclosure for the overall group was 17 years of age, and the mean age of dating for the whole group was 17. The fertility disclosure age of 16 and dating age of 19 was noted for the individual with a homozygous N1303K mutation (N=1). The men with DF508 homozygous mutation (N=5) had a slightly higher fertility disclosure age of 17.8 years with an average dating age of 15.8 and a higher age of fertility disclosure for those who had heterozygote mutations (N=3) in this study at 23.3 years of age and an average dating age of 13.3 years old.

Men reflecting on their adolescent experience showed slight mutational differences, the N1303K individual expressed his stressors were more regarding illness and stigma of having CF. Those homozygous with DF508 mutations reported their stressors within one category: infertility worry (inability to have kids and risk to pass on CF), with more participants expressing concern about worry about the inability to resolve infertility concerns more frequently than their counterparts. Those with compound heterozygous mutations reported decreased time with peers and dates, infertility worry, and stigmatization as compounded concerns.

The average age the males reported that they found out their fertility status was 17 (N=14). When examining those with a homozygous mutation of DF508, the average age of the males who reported finding out their fertility status was 20.8. For those heterozygous with DF508 and another mutation, the average age that they reported finding out about their fertility status was 15. For the individual homozygous for N1303K mutation, fertility status was made know at their age 18.

All participants except one responded with the age that they talked to a healthcare provider about fertility. The average age participants talked with a provider about RSH on their own was 20.1 years. The individual homozygous for N1303K mutation found out at 16 years of age. The individuals homozygous for DF508 found out at a mean age of 17.8 years of age, and individuals who were heterozygotes found out at 23.3 years of age. About their conversations with HCP, the man who was homozygous for N1303K recalled only that his HCP informed him that “I probably do not have a vas deferens but still use condoms.” Those with homozygous DF508 mutations reported being told “nothing” (N=1), or primarily about “fertility status concerns such as only a small percentage of

men with CF could have children but that there were other fathering options” (N=1). Another individual homozygous for DF508 reported, “He never gave me any information on RSH we just had short discussions about it” (infertility) (N=1). “He explained mechanics of why I can’t get a girl pregnant.” Those who were compound heterozygotes were reported to be told “just because I cannot create life does not give you free pass for unsafe sexual practices” (N=1) and that “I had a 15% chance of fathering a child with IVF option” (N=1). Only one individual noted more than one discussion on the topic of RSH with HCPs and no man reported being told all seven topics by their HCP.

The average age of communication for men with HCP without their parent present was 22.2 for males homozygous with DF508, 17.3 for those with heterozygous mutations involving DF508, and 17 for the individual homozygous with N1303K. The average age of this discussion was reported to be 26 years old for those with homozygous DF508 mutation at 26 years of age, 18.6 years of age for the heterozygotes in the study, and 32 years old for the homozygous individual with N1303K.

Extending this concept, 11/16 (69%) men had not been told about ICSI with an equal distribution of men reported that they would like to hear it from a genetic counselor (4/16 men, 25%), pulmonologist (4/16 men, 25%), primary care physician (4/16 men, 25%), and parents (4/16 men, 25%). The individual with N1303K mutation reported he was 32 years old and had never heard of family planning technologies like ICSI. The homozygotes with DF508 mutation had various levels of comfort and discomfort with the use of ICSI to plan families and the heterozygote group all reported being extremely comfortable with the use of ICSI for family planning. The data for RSH timeline for all mutations represented in the study are represented both in Table 2.4 and in Figure 2.3.

Figure 2.3 specifically describes critical RSH events from adolescence characterized in the literature. Here the events and the average ages that the men accomplished those events are listed. Furthermore, in the denoted events of fertility status disclosure and 1st dialogue with an HCP regarding sperm analysis, the average age the event occurred is contrasted with the requested age the men desire these events to happen.

Table 2.4: Age at Critical RSH Events for Males with CF

Diagnosis	Infertility	RSH	Sperm Analysis	ICSI	Dating	Family Mutation
1	18	18	24	0	22	24 DDF508
5	14	15	18	0	8	13 DDF508
13	39	40	39	0	13	35 DDF508
7	20	21	24	0	14	26 DDF508
6	0	0	0	0	14	33 DDF508
1	13	17	18	0	13	20 DDF508
1	14	19	20	0	13	16 DDF508 + 3120 + 1G>A
1	14	16	18	20	14	25 DDF508 + G542X
39	17	20	0	0	16	25 DDF508, UNK
2	18	17	27	0	14	26 N1303K + N1303K
19	0	13	0	0	16	28 UNKNOWN
1	22	25	0	0	15	24 UNKNOWN
4	16	25	25	25	5	25 UNLISTED
8	20	20	25	0	16	21 UNLISTED
2	18	18	0	0	15	28 UNLISTED
1	16	17	0	0	19	19 UNLISTED

2.5 DISCUSSION

To begin the characterization of paternal child sexual communication within genetic conditions, the use of existing RSH literature for men with CF is examined. The current literature discussing RSH needs does not address early learning of the RSH information for men with CF. Furthermore, these previous studies do not consider

mutational spectrum analysis, which is emergent with the advances of genomic medicine and testing in expanded populations and ethnicities.

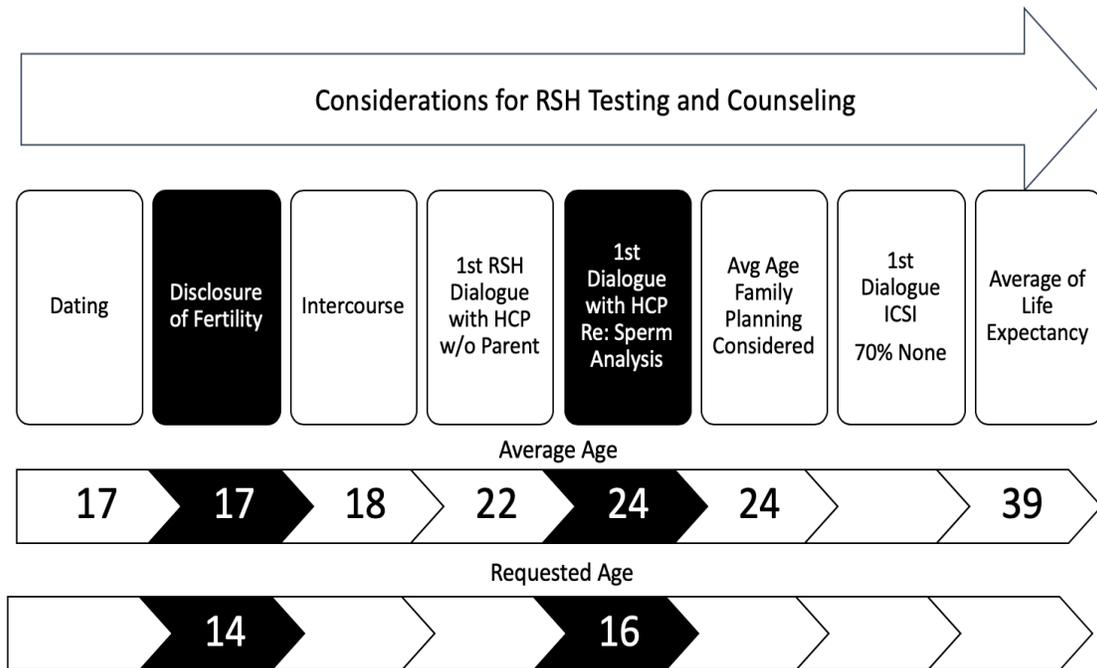


Figure 2.3: Timeline of When Key RSH Events Occurred for Males with CF

Sawyer and colleagues (2005), Rodgers and colleagues (2000), and this study show a lag in RSH service delivery in comparison to CF male preferences persisting over the past 20 years. This study explored early impressions of what constitutes adolescent sexual education that informs RSH as perceived by males with CF and rates father-son communication based on topical concerns. It then teased out the topical concerns that are explored by fathers and those that are explored between the patient and the HCPs, considering that RSH communication and timing of critical RSH information are important to this population due to shortened life span. Based on this preliminary data, consideration of how HCPs, particularly genetic counselors, can facilitate further research and clinical practice that reduces gaps in clinical care are proposed. While RSH studies

for males with CF currently exist, they do not address early learning of the RSH information for men with CF.

2.5.1 Impact of CF on Sexual Education

CF is a multi-system genetic condition and the sexual education needs of the men in this study reflect the same. Their needs, or topics identified, span several domains outside of what is traditionally thought of as sexual education. These topics addressed domains of personal and relational quality of life such as areas of reproductive, sexual, interpersonal, and psychological health, illuminated six adolescent stressors, and highlighted 32 sexual education topics. Also, some sexual education topics were mentioned that are not traditionally thought of as sexual education e.g. medication and nutritional maintenance (that were linked to the ability to be present with people they were dating). This holistic set of issues may necessitate a more integrated approach to male sexual education for parents and HCPs working with this population than may have previously been considered.

Sexual education has both public health implications (initial age of sexual activity, unplanned pregnancies, sexual abuse and STI rates) for HCP and personal implications of well-being for individuals affected. Men with CF have distinct RSH needs stemming from the fact that infertility is associated with 95-98% of males with this condition. This is unique to this adolescent population and a contrasted concern to their male peers who do not have CF.

While this study did not look at the initial age of sexual activity or sexually transmitted infection rates, there were findings of note from a broader public health perspective. For instance, Fair, Griffiths & Osman (2000) suggest that appropriate age to

begin learning about fertility is approximately 14 years old, but disclosure is not happening at this age. A related question sparked by the males' responses became whether early sexual education messages of infertility translated to increased sexual activity and risk for STIs, without appropriate safe sex practices introduced? No conclusions can be drawn from this study but anecdotal reports from the men, demonstrate lessons that can be considered. Ten men reported not hearing any safe sex messages from their fathers and several men described themselves as "promiscuous" before "settling down." More than one male linked that a concurrent message of safe sex along with an explanation of infertility is important. This theme of safe sex education was desired and identified as a significant code for RSH communication with males who are diagnosed with CF by PI as well as NVivo12 software.

The men mentioned that an explanation of their infertility generally included the fact that 95-98% of males with CF cannot get a woman pregnant. For some men, this message was erroneously perceived as an ability to have sex without *any* negative consequences including STIs. Conversely, 2-5% of males in this population are not infertile but it may be perceived, from an adolescent male perspective, that *no* male with CF can get a woman pregnant. This is an erroneous message as well. Hearing or reading that 95-98% of males with CF are infertile, without knowing the details of one's own reproductive capacity, can lead to unintended pregnancy. This was the result of one participant whose encounter of unprotected sex led to an unexpected pregnancy with his partner. He reports his HCP informed him that he was infertile. Sexual education about infertility, the risks of STIs, the use of safe sex, and the potential for pregnancy must be individually counseled.

But what potential impact does CF have on a personal level? Represented in several ways in the table, the men with CF are concerned about fertility and its specifics. The RSH literature for males with CF highlighted that men are typically aware of their fertility status during adolescence and enter relationships with the knowledge or expectations of infertility before entering romantic relationships. This foreknowledge of infertility is incongruent, in some respects, with the preliminary data of this study in that the men were aware of their fertility status in late adolescence with the mean age of fertility disclosure and dating for the overall group at 17 years of age. Some mutational groups were not aware of their specific allelic variants, much less their fertility status until early into their third decade. In more than one male participant, knowing that he was infertile did not necessarily confer clarity as to why he was infertile. With one participant reporting, this was not fully explained to him by a parent or provider until he got engaged which he reported as frustrating. More thorough explanations of fertility at an age congruent with previous RSH CF studies may be needed. The men in this study reported fertility information as the desired part of communication with their father as well as a necessary part of comprehensive information for adolescent males with CF. This may suggest infertility and its implications may need to be staged developmentally over time.

Another personal consideration for males with CF is that they can express concern about their reproductive capacity and how to address or resolve a desire for a family. This was echoed throughout the study by all the men at some point. For instance, one individual reported that “Initially dating was not a problem for me. I could eat a lot without gaining weight and could have sex without fear of getting a girl pregnant, unfortunately, I’m 31 now and want children.” Another individual reported, “I have

convinced myself I didn't want kids but that's not true and I know my wife deeply wants kids too." Though males with CF are living longer there is still a reduced life expectancy. Reconciling infertility with the male's desire for a family appeared a difficult series of choices and life experiences for the men. There were also additional psychosocial concerns that they recalled in adolescence.

All males addressed the psychosocial aspects of their condition. One male stated:

"About 5 years ago after my last relationship, I figured that no one would want to be with me and watch CF slowly take my life and watch me suffer. I figured that enough people would be hurt by my passing already and I shouldn't involve anyone else who would be hurt too. I just stay to myself and keep others at a distance."

One male responded and just said simply that CF impacted him because he "felt self-conscious and unworthy of love." Another reported that he is the second son in his family to have CF and his brother died of CF-related complications while he was an adolescent. While it is noted that other men reported that they did feel emotionally resilient, had come to satisfying conclusions regarding their infertility with their partners and were learning of their worth and value outside of fathering a child, psychological concerns occur often enough for those diagnosed with CF, and their caregivers, that mental health recommendations have been proposed (Quittner, et al., 2016). The sub-questions or sub-themes of the domains listed in Figure 2.2, though addressing a broad range of RSH topics, may possess another unifying theme that may not be initially identified: a theme of "restrictions."

Only four men reported not experiencing CF-related stress during adolescence, the remaining men listed their stressors as the inability to work, the inability to have children, the inability to socialize with or date peers, frequent hospitalizations/or "being

sick”, and an early sense of stigmatization. Some of the questions of reproductive health such as “Why am I infertile?” or “What is my chance to have a child with CF?” are relevant facts that an individual with CF should know. However, this increased knowledge may also be a desire for increased awareness of identity as an individual with a chronic illness. These questions may be a response to a stressor of being “constantly” sick or wondering if their offspring will feel “different” or stigmatized as they do. CF can create stress that impacts an adolescent’s self-concept and may require a family’s understanding of how a sense of stress is created by the restrictions imposed by this illness on adolescent males. Adolescent males’ satisfaction/dissatisfaction with their sexual education may be associated with a sense of restriction/sense of independence.

This PI believes that for males with CF, the condition does have an impact on reproductive and sexual health needs spanning several domains. Sexual education must be thoughtfully layered and medically informed by parents and healthcare providers working in collaboration with one another. Sexual education includes knowledge of a child’s genotype and phenotype, reproductive capacity and sensitivity to adolescent development of a male with CF. Its complexity is best delivered with awareness of its many moving parts.

2.5.2 Father-Son Communication: Primary Resources and Primary Sources

Previous literature indicates that the sexual health of adolescents is shown to be influenced by their parents and is associated with condom use, decreased unprotected sex, and increased protection from HIV and STDs (Flores & Barroso, 2017). Also, while most literature features sexual education as handled by a mother, Kapungu (2010) showed that the number of covered topics is highest between same-gender dyads with fathers

generally communicating with their sons. This study considered sexual education from the standpoint of resource utilization, topics of discussions covered between fathers and sons, who initiates these discussions, and the males' perceptions of the sexual education quality received from fathers both individually and as a collective.

Even though fathers are considered primary sexual educators of their sons (Breuner, et al., 2016; Kapungu, 2010; Wyckoff, 2008), in this preliminary study they were not the primary resource for sexual education. Participants, during their adolescence, reported actively seeking answers to the RSH questions from various resources that were not consistent among the resident pool. Most participants cited multiple sources for their education during the ages of 10-13 years, but during the ages of 10-13, the men still cited their father as one of their primary sources.

While there are few studies on preadolescent RSH communication with parents, sexuality communications between parents and preadolescents are reported in more than one study to be dependent on gender (Wyckoff, 2008). In the Wyckoff (2008) study with preadolescents and their parents it was found that preadolescents are internalizing sexuality communication given by parents and that when this is done before sexual activity and before increased peer influence, family prevention messages can have more impact (Wyckoff, 2008). The inclusion of fathers as a relevant source of information for the males during the ages of 10-13 is congruent with existing literature that fathers may hold slightly more influence over their male preadolescent and early adolescent children in sexual education than later adolescence. If confirmed on a larger scale for males with CF, this PI believes that the data may imply a window of opportunity, early in a child's development where fathers can capitalize on their influence over their son and provide an

early avenue of open communication to areas of comfort between both parties (This is also supported by results in Ballard & Gross, 2009). In typical adolescent sexual education literature, preadolescence can be a time, to begin with, less sensitive topics such as hygiene or body parts, and build knowledge, skills, and confidence to engage their son's rapport for the more comprehensive challenges ahead, e.g. infertility.

The resource elected most by the men for sexual education were friends who were not familiar with CF, personal experience, and their father. However, by the ages of 14-18 years, fathers were no longer considered resources by males with CF to address their son's sexual education needs. During the ages of 14-18 years, the participants reported that their sources of information for sexual education were personal experiences, friends unfamiliar with CF, and internet websites. Fathers and books were lesser utilized resources to learn about dating, sex, and relationships.

This study supports the complexity of sexual education for males with CF. Despite fathers not being used as a primary resource, literature does cite that parents are still the primary educators of their children and sexual attitudes are always transmitted, even if by default or silence. In failing to discuss issues of sexuality early, fathers may neither be identified as the primary source of sexual education nor acknowledged as a resource; but, may still be conferring attitudes and behaviors about how to approach and discuss sex with their inaction (Wycoff 2008).

Both fathers and sons quote personal narratives to negotiate sexual education communication that is congruent with literature (Coakley 2017) but the sources fathers use to educate their sons (faith-based resources and books) and the resources sons use to educate themselves (friends and internet) may interfere with sexual education messaging.

For the adolescent male, the use of the internet may pose some concerns. The accuracy of what is supplied via internet sites may be called into question especially if trusted resources are not utilized and information is pulled from blogs or social media. To counter this concern Coakley and colleagues (2017) suggests practitioners steer families toward reputable information and teach them how to identify subjective instead of objective information.

Males with CF in this study reflected that, as adolescents, they used personal experience to get their reproductive information. Fathers in previous literature reported that personal stories assisted real-life information that some fathers helpful to ease the transition to more graphic or explicit situations. Mostly, personal experience is used to demonstrate what to do or not to do in certain circumstances and to have the benefit of natural conversations that expresses parental values and expectations (Malacane & Beckmeyer, 2016). Having personal experience also allow sons to modify what they are being told. Knowing how to speak effectively with their sons is a crucial part of education. Transitioning parents to be an effective source of sexual education, comprehensively addressing the multiple domains identified by the males in this study encourages them to have proper resources to provide purposeful and intentional messages about CF and RSH.

The use of personal narrative identifies that fathers can draw from their wealth of lived experience to educate their sons. For the father of a son with CF, despite not being affected by the condition, he could refine this technique, inclusive of the impact of CF he observes in his son. Personal narrative is a well-received, well documented and shared strategy in parent-child sexual education. It is simple and effective. In this study, it was

employed in the sexual education of CF males as well. Fathers can grow communication from their sons' early childhood, continue the growth along a trajectory of observing the specific impact of CF on their child, direct needed early interventions and expand communication to be open and, eventually, bi-directional. It may just require tuning into key concerns identified by awareness of their child and initiating relevant elements suggested in this study. It may also require that fathers have necessary supports they need to be successful.

2.5.3 Father-Son Communication: Topics of Discussion

Based on the 32 topics identified in the responses from the adult males, fathers in this study were reported to address almost half of the topics desired by their sons. This occurred to various degrees of comfort within the male recipients. The topics, taken as an aggregate, map out to general sexual education needs and CF-sensitive sexual education needs. CF-sensitive topics identified in this topic were CF-related rejection, self-esteem, belonging, body image, and infertility risk concerns. For general sexual education topics, the fathers covered such things as changes to their body, personal hygiene, "facts of life" (e.g., where do babies come from, how babies are made), how to approach a girl, sexually transmitted diseases, sexual harassment/consensual sex, being in love and handling a breakup. Reviewing the affected males' responses, additional topics such as pornography, drugs, and sex were also discussed. One-third of the fathers addressed safe sex and shared a trend with parents in standard sexual education in discussing general information about the consequences of sex, but they did not address specific fact-based information about such things as intercourse or intimacy (Kapungu, et al., 2010). It is unclear how many of the males in this study who did hear safe sex messaging practiced safe sex during

adolescence. As of 2017, 38-50% of males 15-19 years of age utilized condoms (US Census Data, 2017). The males in this study reported hearing safe sex messages of condom use, abstinence, limiting partners or practicing monogamy, and limiting alcohol and drugs. However, there was insufficient data to assess safe sex use of males.

Over 18 topics related to general sexual education and CF-sensitive sexual education were analyzed with participants reporting various levels of comfort in conversations with fathers about these designated sexual topics. No topic whether comfortable or uncomfortable, received more than 5 votes. The men reported multiple topics that they could comfortably address with their fathers including personal hygiene and how to approach women. A few men reported that they could talk comfortably about being in love and reproduction. Fewer still (<15%) reported comfort approaching their fathers regarding infertility, STI prevention, consensual sex/sexual harassment, body image, and handling a breakup.

There is no real uniformity for the males in this study in what they feel they can comfortably address with their fathers except for what they did not vote for. No one expressed comfort at addressing reproductive anatomy, how their body works, masturbation, intimacy, sexual identity, and talking with their doctors about sexual health. Perhaps the commentary to be made is that without early sexual education maintained in the home, communicating about sex is “hit and miss.” The data shows men have clear ideas of what they want to discuss with their father, but they do not mention the critical RSH information that they deem as necessary to discuss. The question may be asked, “What stops a child from discussing sexual education with their fathers?” Do fathers begin early modeling as initial male authority figures providing a template for

future conversations with subsequent authority figures, such as healthcare providers?

Does unidirectional or poor RSH communication at home get further translate to passive communication or reduced RSH resource access in a clinical setting?

2.5.4 Father-Son Communication: For Better or Worse?

In “21st Century Parent-Child Sex Communication in the United States: a process review,” by Flores and Barroso (2017), the authors reviewed 116 studies addressing parent-child sex communication yielding 44% “mother only” studies, and 7% “father only” studies. They reported that children view their fathers as having inherent authority regarding specific topics that include how men think and they report children would prefer to learn these topics from their fathers. The authors concluded that “the study of fathers’ sex communication is paramount to improve the role of paternal engagement” (p. 544) in practice and in research. This study supports that conclusion. Given the general tone of the codes Figure 2.2 (father, communication, sex, self-esteem, number of sex partners, relationships) about questions relating to fathers and perceived father-son RSH communication, it is suggested that the males desired father-son RSH communication, but by and large viewed their RSH communication as a negative event and process. The RSH communication did not occur or when it did occur was a negative experience. Q17 and Q27 had no or little positive coding associations. Some examples of responses (each from different surveys) include:

“The only comment made left me speechless and was very crude.”

“I think him being too cavalier about certain things might have allowed for the Yolo mentality but my mom’s over-controlling behavior made me feel shame for it.”

There are some parallels to sexual education literature with typical adolescent populations who do not have genetic conditions and findings in this study. Furthermore, insights into these sons' perceptions of their father's communication about sexual education are identified along with the perceived impact of paternal engagement and strong male sentiments about their paternal RSH communication. Males in this study viewed communication with their father negatively. See Figure 2.2. 64% of all recollected father-son communications were coded as "negative" and "very negative" in the sentiment analysis clustered around the topic of father-son RSH communication.

While fathers, in general, covered a broad range of topics (5 themes emerged and are listed in Appendix D and are discussed below), there was little overlap between what the men said they wanted to discuss and what they report their fathers discussed with them. This is borne out in responses regarding their fathers' communication with them. Over 1/3 of males with CF reported there was never any discussion about sex and that their fathers never provided them with any helpful information regarding sex. Three males reported sometimes their fathers' information was helpful but they either yelled at them, discussed "drugs more than sex" or never discussed sex at all as previously mentioned. One male reported his dad was helpful but never discussed infertility, which is something he wanted his father to address. Only one male described his father as being actively engaged in his sexual education. In short, if there was RSH communication from father to son it was not considered education. Males with CF in this study surmised that RSH communication and sexual education conversations were non-specific to their needs and were unsatisfactory.

Also, there did not appear to be an even exchange (bidirectional communication) of sexual education between father and son. This is critical to understand because the sexual health literature reports that adolescents have a perception that initiating conversations and discussing sex may elicit a negative reaction. Sexual education literature states that adolescents feel they may be viewed as being sexually active, face punishment, or be judged. This answers the question as to why males with CF may not initiate sexual education topics of concern. Additionally, men in this study were concerned about paternal reactions they felt they would be unprepared for. Responses from three participants in this study were:

“My father was and is an abusive narcissist...”

“If I was taught anything by him, I would have lived a more dangerous sex life.”

“I would maybe ask him questions, but he would only tell me things he may have encountered and then be disappointed if I did something at a young age that didn't seem appropriate for the age.”

In contrast, parents have admitted that they discussed issues only at their child's initiation and did not talk about sex until asked (Baier, et al., 2008), while some cultures, for example, Muslim mothers have reported they did not think it necessary to initiate conversations about sex and said they were available if their daughters needed to talk (Orgocka, 2004). Sexual education literature also shows that children were unlikely to initiate topics of sexual conversations even if they do have concerns (Flores & Barroso, 2017; Collins, Angera, & Latty, 2008). A lack of communication about sex, therefore, can cause adolescents to feel awkward and anxious to the point of avoiding sexual discussions. Parents, as primary sex educators, have values that are conferred to their

children, even if it is by default (Pagano, et al., 2003). More than one participant in this study reflected this by reporting discomfort that conversations were “too personal” or reported that they were “avoidant like their parents” when it came to addressing issues of RSH.

There are only six times within the participant responses that fathers initiated sexual education. In four of these instances, there was negative messaging. In two accounts, a conversation on sex was initiated due to the son finding his father’s pornography and viewing it. In the other instance, the father discovered that his son had viewed pornography. In both instances the sons reported unidirectional, closed communication, stating “don’t let me catch you with that [his father’s pornography] and “he was yelling at me when he found a porno mag in my room and I just listened.” In two additional accounts, the sons reported sexual education statements vs. conversations reporting that his dad told him to employ safe sex methods and another male respondent recalled his “conversation” was a question, “he asked if I was being responsible, I said yes, and we didn't expand much because I didn't want to talk about it.” In that account, it was unclear why this exchange was deemed unproductive. Was it the son who stopped the RSH communication and was unwilling to talk to his father or was it the father who may have made talking about RSH topics previously uncomfortable? In the literature, some fathers are observed as doing “spot checks” versus two-way conversations (Solebello & Elliott, 2011). A spot check is a sexual education question that makes a reported assumption about the child and perceived as almost rhetorical in nature. Fathers who were reported to have negative communication may have behaviors or communication styles that interfere with their message. Interestingly, a directive

approach about sex with more authoritarian or unidirectional styling does not invite open-ended communication about sex.

In the literature regarding typical adolescent populations, open-ended communication is associated with sexual risk avoidance (Flores & Barroso, 2017). When they do communicate, parents tend to give general communication about the consequences of sex but not specific fact-based information about such things as intercourse or intimacy (Kapungu, et al., 2010) that, with children, and perhaps children with CF, may not be the best course of action as children echo the outright desire for relational aspects of sexual education. Furthermore, while some males in this study mentioned conversations such as disclosure and dealing with emotions, these conversations are overlooked for conversations about actual decision-making and are supported more so than emotions, relationships, and romantic discourse (Stiffler, Stims, & Stern, 2007). Yet fleshing out concepts of the disclosure in the context of relationships, navigating issues of esteem and rejection, and negotiating complex biopsychosocial concerns are what is needed by the male with CF.

Two respondents reported what appears to be inappropriate or ill-timed conversations initiated about sex by their fathers. One adult male reported, “Dad started the conversation about sex and drugs, but it was mostly about drugs.” He went on further to clarify that his father mostly talked to him “about anabolic steroids a lot.” For another respondent, he had a negative reaction to the conversation reporting his father initiated a conversation about sex but “the only comment (he) made left me speechless and was very crude. I had no desire to seek an opinion or guidance from him. I didn’t respect his opinions. But he said, “(essentially) to keep it in my pants’.” Truly, it is hard to know

why there is not more positive CF RSH communication between fathers and their sons, but sexual education literature for adolescent populations without a genetic condition demonstrates that, too often, as evidenced with the men who mentioned incidences with pornography, sexual education in the 21st century remains reactive. By and large, the men had difficulty recalling “conversations” with their dad about sexual communication. Sexual education appears to be of a “one-time” nature, often punctuated with frustration and anxiety. This, too, is a concept paralleled in current sexual education literature of children with non-genetic conditions. (Baier. et al., 2008; Meschke & Dettmer, 2012; Orgocka, 2004).

Fathers of children with CF present with several concerns including being overwhelmed, feeling isolated, experiencing altered family dynamics, the quest for resources, and experiencing financial strain (Shardonofsky, et al., 2019). One cannot underestimate the impact of this condition on fathers and the family system. Genetic diagnoses impact fathers in at least four distinct ways from other family members, according to Rivard & Mastel-Smith (2014); 1) fathers’ reactions to childbirth and disability are different from those of other family members; 2) paternal stressors center on financial issues, family commitments, and feelings of incompetence with the child; 3) fathers’ needs for education about causes of disease, care of the child, and prognosis are not adequately being addressed; and 4) paternal responses, including stress and guilt, have a great impact on the child. Several studies have highlighted these same concerns specific to CF. Therefore, anticipatory oversight regarding patient education, available support resources or informational groups for fathers, written resources or information sent home for family may need to be a considered standard. The perceptions of fathers

are beyond the scope of this study but identifying areas that can be strengthened between father and son is within the scope of this paper and some of the findings suggest this can be completed.

2.5.5 Father-Son Communication: Toward a Better Future

There was partial evidence for greater frequency of sexual communication in non-white/Caucasian vs white/Caucasian households. Two respondents from non-white/Caucasian backgrounds reported important messaging conveyed in communication with their fathers versus white participants, who did not identify that sexual communication was present or positive. This is significant to note that demographic factors such as socio-economic background, ethnicity, gender, and other demographics play a smaller but still notable role (Jermain & Constantine, 2010) in how fathers and sons communicate, about what, and how often.

Although no explicit positive conversations were cited by the men, positive codes emerged suggesting some positive sentiment in messaging. Some men did report positive things that their father specifically did or said that was helpful:

“kept an open mind and tried to answer my questions”

“he assured me (despite CF) that there would be a lot of girls in my life that I would like or would like me and my time would come when it was right for me”

“he always wanted me to be more focused on my studies and not worry about having a serious relationship until I found my direction.”

Each of these statements was self-selected by the participant as important messages to them as a male with CF and address belonging and topics of interest for males with CF. Father and son RSH communication and respect, in these examples were valued and mirrored conclusions that paternal involvement can decrease sense of negative self-

consciousness and provide directive communication that navigates aspects of uncertainty in CF. The topic of father-son RSH demonstrates that fathers want to instill a sense of responsibility so that their sons can learn from their personal stories and be trusted to protect and provide for themselves (Dilorio, et al., 2006).

About the harmful messaging or negative conversations about sex, outside of what has been previously mentioned, one respondent reported that he wanted sexual education to include handling rejection that comes with CF and dating. He reported that his father “never really taught me how to deal with my emotions when someone hurt me by breaking up with me or doing something that hurt me so I would act out impulsively and I would usually get myself into trouble trying to deal with it..” Another respondent reported, my father “would always be inappropriate when referring to any sexual conversations.” While we do not know the context of these respondent’s perceptions of their father, these accounts veer away from the literature reporting parents can be a gateway to positive sexual communication. This is important to note because not all parents will be capable of being healthy sexual educators for their children, so providing avenues where clear reproductive sexual health resources for a male with CF is available becomes even more important. The source of an online adjunct to CF-specific sexual education in middle school or finding reputable sources and modalities to be educated can be affordably done or accessed. One male suggesting online YouTube videos.

Perhaps the most damaging message is that of no message at all. One member reports, “avoiding these (sexual education) topics forced me to be in the dark and look to other people or school classes for answers”; another respondent put that the most harmful message his dad gave during their sexual education was “his lack of communication.”

Few men could not recall any positive messaging given while most men did not identify their father initiating any sexual conversations or providing any safe sex messaging. Likewise, more than one participant in this study reported that sexual discussions were not conversations but precautionary statements that discourage sexual behaviors. In this study, two men addressed the fear that their parents had that they may sleep around or, in one man's concern, that he may be more promiscuous based on his father's concern.

Parents may avoid speaking about sex because they feel that it may give children permission to have sex or send mixed messages (Dilorio, et al., 2006; Meshke & Dettmer 2012). Even more so, pleasure and positive aspects of sex are considered off-limits. From adolescent perspectives while sex communication is essential to prevent risky behavior, scare tactics are ineffective and initiation is difficult. So, while this finding comes from parent-child sexual education from typical adolescent populations, the CF adolescent male has parallel ideas of what he needs and also how he wants it to be relayed. CF males had one set of priorities, their fathers had another. While there may not be an even exchange of sexual education dialogue at the time of this study, there is evidence that communication between both parties is desired. Overall, being a sex educator as a parent can be rewarding but the decisional balance required is not easy.

While they are interested in hearing things from their father, most men felt their father's sexual education could not serve as standalone education. Notably, almost a third of the men reported that sexual education would have been no different if it had been provided by their fathers alone, which may mean that their father's sexual education was noncontributory or quite contributory depending on the perspective.

While there is a growing body of literature regarding father and positive health outcomes with their children, there are at least two instances cited in Coakley, Randolph, Coard, and Ritchwood (2019) that questioned whether fathers who engaged their children in RSH communication, shared developmentally appropriate topics. While developmentally inappropriate messages were referenced by two participants, more men reported decreased quality of sexual education if provided by their father's alone. As more is learned about genetic conditions and sexual education, more resources can be made available to fathers. Likewise, there is promising literature that men desired to be better educators for their children and that fathers of children with genetic conditions, including CF, need extended support and training (Kobylianskii, et al., 2018; Pelentsov, et al., 2015; Shardonofsky, et al., 2019).

The men reported wanting father-son communication to address reproductive, sexual, and relational health. They describe looking to their fathers for guidance in establishing values and addressing recurrence risk, impact to reproductive health, and specific advice on how to deal with partners including having patience and honesty in relationships and not being afraid to show emotion and sexual direction regarding women. The men expressed concern about the impact of CF on sexual performance and felt unprepared in this area. They did express wanting a father to specifically address early adolescent concerns as to whether bathing before and after sex is necessary, risk of illness from sex, sexual side effects of medication, and endurance, pacing, and stamina required for sexual activity. They request of what they report wanting is more explicit from the type of topics being exchanged comfortably between both parties. It is unclear if the men were aware of community resources, such as from Cystic Fibrosis Foundation

centers, which may address some of their sexual education topics. Existing resources are being untapped but there also seems to be a need for more comprehensive resources and resource sites for males with CF entering adulthood. This would be a resource that may assist both father and sons alike during this transitional period. There was no association observed in this study between the rating of the father-son relationship and the positive or negative impact of sexual education with this small sample.

The men's perception of how their father impacted their sexual decisions may offer a final piece to how they qualify their sexual education from their fathers as the majority of men can cite some impact that their father's values and beliefs had on their decision making for better or for worse. Fathers are one of many resources that adolescents use to become sexually educated and their communication, when strengthened, is powerful. The needs of the males who responded to this questionnaire were congruent to the sexual health needs of typical adolescent populations, but they expressed the additional need for more directed information to address their unique reproductive and sexual health concerns. Sexual education cannot be treated as CF sexual education. However, fathers may need some additional awareness of what topics and what resources to address. They also need to know how best to reinforce age appropriate RSH messaging when speaking with their male sons with CF both, one on one, as a parental unit, or as part of a conjoint effort with a HCP.

2.5.6 RSH Communication with Health Care Providers

2.5.6.1 Males with CF Have Misconceptions About RSH

Since 2000, the literature has documented that men with CF have expectations for normal sex relationships and hope for children. They also express increased sexual

education at earlier ages. Men with CF have documented an average age of sexual intercourse at 17.9 years old (Sawyer, et al., 2005), requested semen analysis at age 17 years, and reported a 38% use of condoms (Sawyer, et al., 2005) Also discrepancies have previously been identified in sexual education knowledge. Popli and colleagues (2009), and Thickett and colleagues (2001) reported that men with CF did not feel they needed to use a condom, largely because they confused infertility with erectile dysfunction. And in the Sawyer, and colleagues (2005; 2009) studies, males with CF overestimated their risk to have a child with CF when considering IVF. Males with CF were also unfamiliar with the concept of small volume ejaculations in men with CF (Popli, Bourke, & Stewart, 2009). In this study, we explored if RSH needs differ across mutational types to see if specific sexual education needs may relate to mutational class but there was not enough participants and mutational class distinction for this analysis to be relevant at this time.

In comparing what is available in the literature and what was demonstrated in this study, the preferred age and actual age of RSH information delivery revealed interesting lapses, worthy of investigation. For instance, considering general conversations with an HCP without parental presence demonstrated a difference. The average age participants talked with a provider about by RSH independently, was 22.1 years old for those homozygous with DF508 and 17 years old for those with other mutational subtypes. Adult males in the Fair and colleagues (2000) study desired these conversations earlier, before the age of 16. In this study, no mutational group discussed RSH alone with an HCP before the age of 16 years. Even if these men were not infertile, they would still benefit from sexual education at a younger age to process, and even develop RSH questions. With this genetic condition, there may be ignorance about sexual lives of

adolescents with CF. Taking into account variable disease presentations, the average age of sexual intercourse for a male with CF was 17.9 years old. Lack of conversations about RSH put males with CF at a disadvantage for addressing important aspects of sexual health and well-being, after the average age of their first sexual encounter.

In considering disclosure of fertility status, in this research, the average age of learning about fertility status was 17 years old. However, it is suggested that the appropriate age to begin learning about fertility is approximately 14 years old for the male with CF (Fair, et al., 2000). Also, close to half the participants reported they learned about their fertility status “on their own” and there is almost a 1.5x increase in the men who found out their fertility status “on their own” than in a Rodgers and colleagues’ study (2000) that occurred 20 years ago.

While previous studies list HCPs as preferred sources of disclosure, this preliminary study found that the men preferred both parents to be the one to disclose their fertility status. However, it was also suggested in this study that the fertility status disclosure conversation would be welcomed if given by either parent or at least one HCP. Based on the small sample represented in this study, individuals who had heterozygous mutations found out their fertility status before those with a homozygous mutation. Yet less than a quarter of the total individuals in this study reported that they heard their fertility status from their preferred sources. Men with CF want adequate discussions with an HCP about fertility and they want to hear from their preferred source.

When discussing the communication from their health providers during adolescence, the message cited as “most helpful”, was hearing about “fathering solutions,” including IVF. Males with CF were receptive to discussions about IVF,

testosterone supplementation, adoption, and fostering. This is important because, in the data of this study, the men considered wanting to “have a family” as early as 13 years old, but with an average age of 25 years old.

Multiple topics were covered by HCPs such as infertility, specifically the difficulty/inability to have children and the “physical issue” (assuming this refers to physiological reasons why participants may not be able to have children). Providers reinforced the importance of safe sex and the possibility of IVF. However, the men, as an aggregate group, reported less than a quarter of the topics covered, though to a varying degree. Some providers were perceived to follow up facts with supportive information to a greater extent than others. For instance, the men clarified that they had discussions that informed them that they were infertile but the physiology of why infertility was occurring was not explained. One male reported the opposite, that his provider discussed inheritance, the physiology of his infertility, and safe sex options. He reported this was helpful as he didn’t want any child going through what he went through.

Infertility and the reasons why some men are fertile (and some are infertile), though explained, may be difficult to comprehend as reported by one participant who had a college degree. Yet, another male reported that his doctor just inquired if he was having sex and never explored or conducted an RSH conversation further. Partial messages with no follow up can inhibit comprehension and interpretation. HCP are encouraged to personalize care for CF male patients based on successive conversations regarding infertility and sperm analysis rather than a “one-time” general information session (Frayman, et al., 2008; Rodgers, Baldwin, & Knox, 2000). Overall conversations with HCP were seen as providing helpful and valuable information with only one person who

perceived that his fertility status was withheld as a form of birth control and three men who reported that their provider discussed the “heavy price tag” and “expense” of IVF. The men found these messages were conveyed with negative connotations, one man reported his HCP told him, “if you ever want kids it will come with a heavy price tag.”

When considering the conversations had with their providers, half of the men reported hearing a message of utilizing safe sex practices. This appeared to fill a gap that was not addressed by the participants’ fathers. HCPs catch a subset of males who may have limited information and provide them with meaningful RSH information they remember.

2.5.6.2 Family Planning Technologies

Three communication touchpoints were addressed in this study. Speaking with their HCP about their fertility status, semen analysis, and family planning technologies. The men identified several HCP including pulmonologists, geneticists, and genetic counselors who coordinate their care. Men with CF are living longer, and technological advances now permit men with CF to have children. The desire for a family exists (Fair, et al., 2000; Sawyer, et al., 1998; Thickett, et al., 2001).

The discussion age of semen analysis ranged from 18-32 years old, while six men reported that they have not had any discussion regarding semen analysis. In 2005, Sawyer and colleagues reported men with CF have been advocating for semen analysis before the age of 20 for adequate family planning consideration. Most men had never been told about ICSI but wanted to learn more from their genetic counselor, pulmonologists, primary care physician, and/or parents. The homozygous DF508 mutational group, who were aware of ICSI, presented with a range of comfort levels with the use of ICSI to plan

families. The compound heterozygote groups, who were aware of ICSI, all reported being extremely comfortable with the use of ICSI for family planning and the homozygote N1303K individual reported he had no awareness of ICSI and as such couldn't evaluate his comfort level.

Little is known regarding initiating a conversation between CF male patients and HCPs regarding family planning. HCPs appears reluctant to provide more family planning information. It has been introduced in the literature that ICSI could pose some ethical concerns or disequilibrium for providers who provide carrier screening for identifying pregnancies at risk for CF would now have to counsel regarding family planning technology to have a child with CF through ICSI (Sawyer, et al., 2005). In addition it is also this PI's position that HCPs are not a homogeneous group but rather multiple providers who may concur that this conversation of ICSI may be best had by a reproductive endocrinologist. Family planning and family planning technologies are still a topic that adult males do not know how to bring up but want to know more. If healthcare providers are open or provide printed resources that address these conversations, their patients may receive more beneficial information at earlier ages.

There were several psychosocial considerations listed by these men regarding family planning. Four men were concerned at the "cost of a chance to create life." While most men were married or in a committed relationship, they did express having to have long conversations about risks with their dating partners who were considering families. One man reported "I had the choice to use a medical procedure to collect sperm or use donor. I just could not bring myself to use mine and then have my kids be a carrier and risk their kids with the same or with CF, too much guilt to do." The men weighed

considerations like their reduced life expectancy with one reporting, “because of the question of life expectancy, I'm not sure if I want to have children with my wife for fear of dying too soon and leaving my wife with the responsibility. Another stated, “I am not sure I want to do IVF and potentially pass on the CF gene.” The men are resolving their family planning needs in various ways, but the question becomes if they can obtain the information sooner by HCP, can they resolve these concerns sooner?

This research demonstrates a CF RSH timeline that can be quite significant in context. Like a woman whose “biological clock” is ticking, the men experience quite a different “biological clock” due to intra- and interpersonal pressures of infertility and shortened life span. It appears as if limited conversations or no conversations of critical importance could put them at a disadvantage of what to ask, when to ask, or who to ask further questions due to lack of knowledge. Limited cases may also subtly reinforce a lack of personal agency in health care decisions. In cases where they are not being provided follow up information about something as central as reproductive options or not hearing about reproductive options from their preferred source, HCPs can unwittingly communicate a subtle, though unintended, disrespect of the men’s life course and reproductive needs, almost an unnecessary reproductive injustice. Currently, annual recommendations for mental health evaluations were made by Quittner and colleagues (2016), due to caregiver and patient stress inherent with CF care. However, what proportion of mental health can be alleviated by facilitating earlier RSH communications and initiating more robust conversations about family planning technologies as an option for males with obstructive infertility, most of whom, express a desire to have a family? Pulmonologists and PCPs may not be the specialists to convey this information, but they

may be key players, along with GCs, to refer males with CF to a reproductive endocrinologist or a urologist when indicated.

2.5.7 Genomic Testing, Precision Medicine, and Genetic Counseling Implications

In an era of genomic and precision care medicine, particularly across diverse populations, we are discovering new facts about CF. Approximately 70% of individuals who test positive for CF have the DF508 mutation and literature accurately reports occurrence mainly in Caucasian or white populations. In the past, we have not always had the technology to look for and detect the remaining 30% of mutations. However, as technology has evolved and access has been expanded globally, we now see that what was once considered a monogenic disease primarily in one population is heterogeneous within Caucasian populations and those of other ethnicities. Technology is detecting heterozygous mutation of DF508 mutation along with mutations such as G542X, N1303K, 3120 +1G>A, or homozygous mutations in these rare mutations that are represented in our studies.

This is significant because some of the less-common mutations are associated with variable presentation, including later diagnosis, fewer hospitalizations, increased sperm production, or life expectancy. Keeping abreast of these nuances when counseling populations within a genetic condition, impact what and how an individual male may perceive the economic, psychosocial, and physical complexities associated with this genetic condition and certainly impacts the reproductive and sexual health messaging to males with CF. Genetic counselors have the unique expertise to be able to describe specific genotype-phenotype correlations to patients.

The sample size of this study is too small to make broad generalizations but having participants with rare mutations allowed for anecdotal comparisons of how different genotypic expressions could relate to educational needs. In this study, men reflecting on their adolescent experience showed slight mutational differences; the homozygous N1303K individual expressed his stressors were more regarding illness and stigma of having CF. Those homozygous with DF508 mutations reported their stressors within one category: infertility worry (inability to have kids and risk to pass on CF). These individuals more often expressed concern about their worry about the inability to resolve infertility concerns compared to their counterparts. Those with compound heterozygous mutations reported decreased time with peers and dates, infertility worry, and stigmatization as compounded concerns.

Communication with HCP about RSH without a parent present occurred later for those with a DF508 homozygous mutation than for the other group by five years. Discussion with HCP regarding semen analysis occurred for the heterozygote group before the preferred age of 20 but occurred 6-12 years later for the homozygous DF508 and N1303K individuals. Finally, the heterozygotes reported uniform comfort with the use of family planning technologies while those homozygous for DF508 expressed varying levels of both comfort and discomfort and the N1303K individual reported never having heard of family planning technology or ICSI. These differences could reflect mutational differences in sexual education but could also reflect factors such as the HCP-patient relationship, the patient frame of mind, various degrees of health, or access to healthcare that could differ among each participant.

Despite suggestions in the literature, there are currently no formal guidelines for physicians regarding sexual and reproductive healthcare for men with CF. Sawyer and colleagues (2008) reported seven points of information for men with CF: that 95-98% of men with CF are infertile; that infertility does not mean impotence; that spermatogenesis is likely to be normal; that ejaculatory fluid will not contain sperm because of bilateral absence of the vas deferens; that ejaculatory fluid is of low volume; that sperm aspiration and intracytoplasmic sperm injection can result in successful pregnancy; and that genetic counseling is available that are points that would be welcomed by participants of this preliminary study. Since there is no formal CF RSH guideline of care for genetic counseling, it may help note how this preliminary study reaffirms previously established findings and consider it in informing more comprehensive care for males with CF. Therefore, maybe there is benefit in experts considering adding specific recommendations and proposing RSH guidelines for those adult males diagnosed with CF.

Data in this present study can indicate opportunities for potential interventions of males with CF at critical times during their reproductive and sexual development, specifically in adolescence and adulthood. This can be accomplished on both a provider to patient interaction level and system-wide level as HCP coordinate care with each other, families, and patient partners.

Out of the 32 topical areas that are addressed, fathers cover less than half of the topics, and HCP cover less than a quarter. The training and background of GCs allow coverage and reinforcement of most topics. Through combined paternal, HCP, and GC involvement, over three-quarters of the topics could be addressed in adolescent

transitional care. Nine topics are best handled in a home setting and are also topics unique to father-son communication. In addition, some sexual education classes and personal experience can reinforce remaining or previously covered concerns.

Primarily GCs can assist in helping the patient to understand the GC role and their differentiation from other providers. A GC has unique training to help identify early stressors and, in some cases, provide anticipatory guidance of these stressors depending on the age of the patient. They can also discuss a child's unique experience related to having a genetic condition and can provide condition-specific educational and support resources. GCs can provide faith-based Cystic Fibrosis Foundation resources and recommend parental/peer mentoring programs for both parent and child.

GCs can serve as a provider of reliable knowledge about features of CF including reproductive health concerns mentioned by men. GCs can explain the physical features of CF, including the cause of male infertility, and the genetic basis of the condition, including the risk of recurrence. GCs can address myths regarding CF and address safe sex messages. Though they may not directly provide sexual education, they can provide parents with anticipatory guidance, trusted internet sources, and help facilitate parent-child communication.

While there are some topic areas, such as sexual performance and relationships that may fall outside of the scope of genetic counseling, GCs can provide anticipatory guidance about how sexual and reproductive health may be impacted both positively and negatively and provide supportive counseling for psychological concerns commensurate with their degree and ability. They are also able to make referrals to mental health providers more difficult psychosocial concerns and can provide appropriate follow, as

recommended by Quittner and colleagues (2016) for caregivers and patients due to the complex nature of the disorder and its impact on family life.

As it relates to interdisciplinary care, GCs can coordinate efforts by CF care teams with patients. They can help coordinate care with RSH providers to address timing concerns for RSH communication. GCs can educate patients generally about what ICSI is, assess if a patient is interested in learning more about this technique and provide necessary recommendations for planning couples wishing to seek more detailed information. GCs tend to have more time allotted for detailed conversations with patients compared to typical HCP, can use this time to address subjects of a sensitive nature. Though not typically used to provide specific CF care for adolescents, this preliminary research may suggest that an appointment with a genetic counselor during adolescence and young adulthood could alleviate some of the misconceptions, stressors, and ignorance in some topic areas and result in more positive health outcomes for this population.

2.6 LIMITATIONS

Despite its strengths, this study has several limitations that include sample size implications, lack of other studies in this area, and definition of paternity. Regarding sample size when investigating patterns across study participants, there are no current guidelines and tools to determine an adequately powered study when conducting thematic analysis. While there are tools with quantitative and qualitative inputs exist and suggest a recommended sample size, all such papers reviewed also submit that some such input for thematic studies cannot be applied prospectively (Fugard, & Potts, 2015; Malterud, Siersma, & Guassora, 2016; Vasileiou, et al., 2018). Such was the case in this study

where, for example, one suggested input for calculation should be the expected population theme prevalence of the least prevalent theme, derived from prior knowledge. To the knowledge of the PI, after the literature review, currently, no such prevalence exists, averting conclusive sample size and power analysis. In part no prevalence may exist due to the absence of other studies which means it is unclear how representative these findings are and how to gauge the meaning of these results.

First, this questionnaire is based on a small sample size and did not specifically probe for experiences of all males with CF so the results are likely on a conservative estimate of actual experiences of males with CF and cannot be extrapolated to general CF male population. The study has not captured or wholly encompassed all experiences of males with CF retrospectively considering RSH messages from their fathers during their adolescence. Nevertheless, it is notable that profound experiences emerged, even in the absence of specific probes indicating that they are significant among this population.

Second, although the present cohort of respondents contained many patients who reported having a non DF508, this is not consistent with published mutational spectrum prevalence data. The PI cannot determine how the type of mutation and hence, the severity of symptoms, affect the current findings.

The present results are qualitative and cannot be used to test hypotheses formally. Although qualitative data allows for the exploration of themes and helps to generate hypotheses for further and more robust investigation, it should be noted that the present sample was incompletely representative and that limits generalizability.

One major implication of the present study is that males with CF who are homosexual may experience reflections on and outcomes from traditional RSH sexual education from their fathers differently from males with CF who identify as heterosexual.

Also, because the majority of participants were recruited from a social media health forum, this may impact the type of messages coded given the population of males with CF who participated may be unique to individuals who are inclined to participate or who have the resources to participate in online social medial health forums.

A small pool of participants was available for this study and there is likely response bias. These participants were not screened for depression or anxiety and complete family situations were unknown, so it is uncertain as to whether psychosocial stress may be compounded in this population. The study involves retrospection by adult males that involves limitations in understanding their fathers' motivations or inaccurate recall of contextual details.

Also, the study did not inquire about paternal communication outside of a biological family or assess maternal contributions.

2.7 FUTURE DIRECTIONS

Future research should incorporate preliminary data to be used to obtain a larger sample size in addition to interview questions targeted for a qualitative study. These changes could better clarify father-son health communication and provide results that could be better extrapolated. Obtaining comparative perspectives of adult males with CF and their fathers or a comparative father group to answer the same or an extended questionnaire could yield a comparative study of fathers' perspectives and provide a balanced understanding of early and late adolescence sexual health. This could also

enhance findings regarding paternal engagement. Though difficult to achieve, obtaining current adolescent perspectives at the time of the study could remove the memory bias present in retrospective studies. Finally, completing this study with a targeted rare group mutational population includes a more diverse ethnic sample that could also identify if true mutational or phenotypic differences can translate to a personalized sexual education or if the mutational differences found in this study are confined to this study.

This study illuminates the need for applied research focusing on policy and practice. Follow up/transitional genetic counseling with adolescent CF males and their families should focus on session interventions and printed resources that facilitate appropriately timed RSH information and testing for the family system, identifying psychosocial stressors that may warrant further attention and guiding informational needs to trusted CF RSH resources for both parents and children. This can improve the ability of males with CF to receive appropriately timed RSH education and help parents, specifically fathers, be able to address the unique needs of their male children affected with CF. Using more tailored approaches could effectively increase sentiment and improve RSH parent-HCP communication. This, in turn, could intersect genetic counseling with Healthy People 2020 goals focusing on RSH to eliminating health disparities, reducing rates of infectious diseases and infertility, and increasing educational attainment, career opportunities, and financial stability (DHHS, 2020). Improving individual reproductive health builds on the foundation of what has been established physically, biologically, and socially in childhood and adolescence.

CHAPTER 3: CONCLUSIONS

In conclusion, this research proposal catalogs elements of adolescent sexual education for males and identifies early RSH communication needs of adolescent males with CF retrospectively through 16 adult male participants. It uses current parent child sexual education literature to draw comparisons and CF-specific contrasts.

This preliminary study suggests that fathers are both active and passive educators of their son's sexual education capable of covering a wide range of topics. Through increased awareness and supportive resources, they may provide significant aspects of their son's care resulting in increased positive sexual health outcomes and well-being. However, current father-son communication and provider-patient communication combined does not appear comprehensive enough to adequately address the unique reproductive and sexual health needs of males with CF, across subtle mutational RSH differences between traditional and rarer mutations and the males exhibit significant negative sentiment over their RSH communication with their fathers.

This study also suggests that males with CF have an expectation of normal sexual health and hope of a family based on recent advances in technology. Reproductive and sexual health needs of males with CF are well characterized to increase sexual health, positive sexual identity, and expand family planning options that suitably address obstructive infertility often associated with this condition. With a shortened lifespan for CF males, the timing of RSH communication matters. This study suggests that

developmentally timed and specialized RSH education provided by genetic counselors can introduce or reinforce needed RSH messaging during adolescence and early adulthood. GC efforts combined with existing father-son communication and coordinated HCP care can provide additional coverage, depth, and exposure to RSH topics addressed in this study. Working together HCPs and families of males with CF can spotlight family planning technologies earlier and facilitate the resolution of infertility worry expressed by males with CF.

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APPENDIX A: CF MALE ONLINE QUESTIONNAIRE

Thank you for beginning this survey. The goal of this research study is to identify what information males with Cystic fibrosis are learning, find out how helpful the information is that they are receiving and hear what males and families most want to know about Cystic fibrosis and sexual education. Ultimately, we want to equip families and providers with the written resources they need to approach reproductive and sexual health for males with CF at age appropriate levels. In this survey you will be asked questions regarding what information you received about reproductive health, reproductive options and sexual health. There are no right or written answers, what is important are your opinions. Most interviewees find the questions interesting. Participating in the survey serves as your consent to participate in this research study which is approved by the University of South Carolina Institutional Review Board. Your participation in this research is completely voluntary and you may choose to withdraw from the research at any time. The information you provide will be kept confidential. In return for your time and effort you will receive a \$20 Visa Gift Card for participation in this study. If you have any questions about this research study, please contact Dianna Sanderson at dianna.sanderson@uscmcd.edu.

- Q1. At what age were you first aware that you had Cystic fibrosis (CF)
- Q2. Do you know your CF mutations? Yes (1) No (2) Skip To: Q3 If Do you know your CF mutations? = Yes. Skip To: Q4 If Do you know your CF mutations? = No
- Q3. What are your CF mutations?
- Q4. How would you describe your relationship with your mother? Very Distant (1) Somewhat distant (2) Neutral (3) Somewhat Close (4) Very Close (5)
- Q5. How would you describe your relationship with your father? Very Distant (1) Somewhat distant (2) Neutral (3) Somewhat Close (4) Very Close (5)
- Q6. Did you have siblings in your home who influenced your understanding of dating, relationships and sex? Yes (4) No (5). Skip To Q7 If: Did you have siblings in your home who influenced your understanding of dating, relationships and ... = Yes. Skip To Q8 If: Did you have siblings in your home who influenced your understanding of dating, relationships and ... = No
- Q7. If you did have siblings that influenced you, please state their relationship to you and age. For example, 2 brother ages 14 and 20. 1 stepsister 16 years old
- Q8. How many hospitalizations have you had in the past year? Please drag the circle across to adjust the numbers.

- Q9. Were there any stressors or benefits you experienced in middle or high school as a young person growing up with CF?
- Q10. Does CF have an impact on your ability to date? A great deal (14) A lot (15) A moderate amount (16) A little (17) None at all (18)
- Q11. Does CF have an impact on your ability to have children? A great deal (11) A lot (12) A moderate amount (13) A little (14) None at all (15)
- Q12. What impact did CF have on your ability to date or have children?
- Q13. Did your perception of how CF has impacted your ability to date or have children changed over time? You can clarify anything needed in the blank box provided. Yes (23) No (24)
- Q14. During the ages of 10-13 (approximately 4th-8th grade), how did you learn about dating, sex and relationships? Please check all that apply. Father (23) Mother (24) Sister(s) (2) Brother(s) (26) Friends who were familiar with CF (32) Friends Unfamiliar with CF (28) Books (29) Sexual Education Classes (30) Internet websites (31) Personal Experience (32) Other Family or Community Member (33)
- Q15. During the ages of 14-18 (approximately 9th -12th grade), how did you learn about dating, sex and relationships? Please check all that apply. Father (11) Mother (12) Sister(s) (13) Brother(s) (14) Friends who were familiar with CF (15) Friends Unfamiliar with CF (16) Books (17) Sexual Education Classes (18) Internet websites (19) Personal Experience (20) Other Family or Community Member (21) Other (22)
- Q16. Which three resources did you trust the most to give you information about dating, relationships and sex? Check all three boxes that apply: Father (1) Mother (2) Sister(s) (3) Brother(s) (4) Friends (5) People you dated (6) Online resources (7) Book/Magazine (8) Other (9)
- Q17. In your opinion, how would your sexual education have been different if it was only provided by your father?
- Q18. Did your father provide helpful information regarding sex, dating and relationships between the ages of 10-18 (middle and high school)? Always (57) Most of the time (58) About half the time (59) Sometimes (60) Never (61)
- Q19. Tell me about a time you talked with your father about sex? You may state something like who started the conversation and your general response to what he said.
- Q20. What safe sex practices did your father address? None, it was not discussed (1) Abstinence (2) Delay Sex, "wait until..." (3) Condoms (4) Be careful with drugs and alcohol (5) Practice monogamy/keep partners to a minimum (6) Other (7)
- Q21. What specific resources did your father use to assist him in communicating with you about dating, relationship and sex education? Check all that apply. Printed Book (3) Printed Magazine (4) Internet website (5) Supportive aid to show what he was discussing examples: condom & handout (6) Faith/Religion based resource (7) Personal stories (8) Another Family/Community Member (9) Community/Faith based class(es) (10) Other (12)

- Q22. What relationship, dating and sex related topics did you feel comfortable talking about with your father. Check all that apply. Changes to Your Body (3348) Personal Hygiene or Care (example; bathe, wear deodorant, etc) (3349) Parts of the Body/Reproductive Anatomy (3350) How your body works (3351) "Facts of Life" (example: how babies are made, where they come from, etc) (3352) How to talk to/approach someone you like (3353) Different Degrees of Intimacy (3354) Masturbation (3355) Disclosure of CF to a Partner (3356) Infertility Risk (3357) Sexually Transmitted Infections/Disease Prevention (3358) Consensual Sex/Sexual Harassment (3359) Sexual Identity (3360) Talking to Doctors about sexual health (3361) Body Image (3362) Rejection, self-esteem and/or belonging (3363) Being in Love (3364) Handling a breakup (3365) Other (3366)
- Q23. What relationship, dating and sex related topics did you feel uncomfortable talking about with your father. Check all that apply. Changes to Your Body (4) Personal Hygiene or Care (example; bathe, wear deodorant, etc) (5) Parts of the Body/Reproductive Anatomy (6) How your body works (7) "Facts of Life" example: how babies are made, where they come from, etc) (8) How to talk to/approach someone you like (9) Different Degrees of Intimacy (10) Talking to What specific resources did your father use to assist him in communicating with you about dating, relationship and sex education? Check all that apply. Printed Book (3) Printed Magazine (4) Internet website (5) Supportive aid to show what he was discussing examples: condom & handout (6) Faith/Religion based resource (7) Personal stories (8) Another Family/Community Member (9) Community/Faith based class(es) (10) Other (12)
- Q24. Why did you feel comfortable/uncomfortable with your father addressing the topics you selected?
- Q25. Did you feel your father was capable of explaining the answers to questions that you asked him about dating, relationships and sex? Yes (1) No (2)
- Q26. What did your father say or do that was most helpful in your talk(s) about dating, relationships and sex?
- Q27. What did your father say or do that was not helpful in your talk(s) about dating, relationships and sex?
- Q28. What relationship, dating and sex related information did you most want to know in middle and high school?
- Q29. What were your biggest worries surrounding sexual education in middle and high school?
- Q30. How did conversations with your father, regarding sexual education, change as you got older? Much better (1) Moderately better (2) Slightly better (3) About the same (4) Slightly worse (5)
- Q31. What ideas about sexuality do you feel should be uniquely included in father-son communication about sex for a child with CF?
- Q32. If you could create sex education resource to meet the needs of parents who have sons with CF, what would it be? What would be included in the resource?
- Q33. Were there any sexual myths that hindered your ability to date or engage with people you wanted to date?
- Q34. How did your father's personal beliefs and values impact your decisions regarding sex, dating and relationships?

- Q35. Would you support a program that engaged parents of young children on how to talk to children about sexuality? Yes (1) No (2)
- Q36. At what age did someone disclose to you that you may be or are infertile? Please drag the circle across to adjust the number.
- Q37. Who first disclosed your fertility (your ability to get a woman pregnant) status to you? Geneticist/ Genetics Doctor (1) Pulmonologist (2) Genetic Counselor (14) Nurse Practitioner (15) Masturbation (11) Disclosure of CF to a Partner (12) Infertility Risk (13) Sexually Transmitted Infections/ Disease Prevention (14) Consensual Sex/ Sexual Harassment (15) Sexual Identity (19) Mother (16) Father (17) Both Parents (18) No one told me I found out on my own (21) I don't remember (19) Other (20)
- Q38. In your opinion whose responsibility is it to discuss a male's fertility status if he has been diagnosed with CF? Primary Care Physician (1) Pulmonologist (2) Genetic Counselor (3) Nurse Practitioner (4) Mother (5) Father (6) Both Parents (7) Other (8)
- Q39. At what age did you talk with a CF healthcare team member about your reproductive or sexual health without a parent present? Please drag the circle across to adjust the number.
- Q40. What information did you receive about your fertility (your ability to get a woman pregnant) and reproductive health that you found particularly helpful at that time?
- Q41. What information did you receive about your fertility (your ability to get a woman pregnant) and reproductive health that you found harmful at that time?
- Q42. What was the take home message about your reproductive and sexual health from your providers when you were in middle/high school?
- Q43. Have your CF HCP ever discussed a sperm analysis with you? Yes (1) No (2) Skip To Q44 If: Have your CF HCP ever discussed a sperm analysis with you? = Yes Skip To Q46 If: Have your CF HCP ever discussed a sperm analysis with you? = No
- Q44. Did your CF HCP ever discuss why a sperm analysis is important medical information to know? Yes (20) Maybe (21) No (22)
- Q45. Please state at what age a sperm analysis was first addressed with you. Please drag the circle across to adjust the number.
- Q46. Has your CF healthcare team ever discussed family planning technology like intracytoplasmic sperm injection (ICSI) with you? If so, please enter at what age do you recall this conversation occurred in the available box? Yes (23) No (24)
- Q47. In your opinion whose responsibility is it to discuss family planning technology like intracytoplasmic sperm injection with males who have been diagnosed with CF? Primary Care Physician (1) Pulmonologist (2) Genetic Counselor (3) Nurse Practitioner (4) Mother (5) Father (6) Both Parents (7)
- Q48. When thinking about whether you want/wanted children, how comfortable do you feel about using family planning technologies like ICSI to plan for a family of your own? Extremely comfortable (21) Somewhat comfortable (22) Neither comfortable nor uncomfortable (23) Somewhat uncomfortable (24) Extremely uncomfortable (25) I need more information before deciding my comfort level (26) I was never told about family planning technologies like ICSI (32)

- Q49. At what age did you begin dating? Please drag the circle across to adjust the number
- Q50. At what age did you begin thinking about having a family? Please drag the circle across to adjust the number.
- Q51. How old are you? Please drag the circle across to adjust the number.
- Q52. Where did you hear about this survey? MUSC (1) CF Clinic Chapel Hill NC (2) Jackson Memorial Health FL (3) University of Miami (4) Piper's Angels (5) Facebook (6) Other (7)
- Q53. What is your highest level of education? Less than high school (1) High school graduate (2) Some college (3) 2-year degree (4) 4-year degree (5) Professional degree (6) Doctorate (7)
- Q54. How do you describe your racial or ethnic background? White (1) Black or African American (2) American Indian or Alaska Native (3) Asian (4) Native Hawaiian or Pacific Islander (5) Bi-racial (6) Multi-racial (7) Other (8)
- Q55. What is your current relationship status? Married (6) Widowed (7) Divorced (8) Separated (9) Single (10) In a committed relationship (11)
- Q56. How may we contact you if we have questions regarding your responses? Check all that may apply. Phone (1) Email (2) Please do not contact me to clarify responses (4)
- Q57. Thank you for your time and for completing this survey. Please provide your contact information so we may verify where to send your gift card. Phone (1) Address (2) Email (3) Please do not send me a gift card (4)

APPENDIX B: REQUEST TO WITHDRAW FROM A RESEARCH STUDY

Request to Withdraw from a Research Study
(Please fill out all sections of the letter)

Name of Principal Investigator:

Title of Study:

IRB ID #:

(ID # can be found in the top right hand corner of your consent form)

I, _____ want to end my participation in this study.
(Name of Participant)

Ending my participation means:

- I will no longer be contacted about this research study unless I need to be notified of a safety concern.
- Information about me, including my health information, will no longer be collected.

I understand that any data collected as part of my participation in the study will remain as part of the study records and cannot be removed.

Signature of Participant/Date

Date of Birth of Participant

APPENDIX C: PARTICIPATION TO CONSENT

Project Title: Checkmate: Father son communication regarding reproduction in children with Cystic fibrosis

Dear Potential Participant:

My name is Dianna Sanderson and I am a graduate student in the School of Medicine Genetic Counseling Program at the University of South Carolina. You are invited to participate in a graduate research study focused on father-son conversations about dating, relationships, and reproductive options for males with Cystic fibrosis (CF).

INFORMATION AND PURPOSE:

The goal of this research is to identify what information males with Cystic fibrosis are receiving, find out how helpful the information is and hear what males and families most want to know about Cystic fibrosis and sexual education. Ultimately, we want to equip families and providers with the written resources they need to speak with their children at age appropriate levels in these areas.

I am interested in hearing from males who are over the age of 18 and are diagnosed with Cystic fibrosis and from fathers who have a son, ages 10-18, diagnosed with Cystic fibrosis. I would like to extend an invitation to you to participate in this research.

YOUR PARTICIPATION:

Participants are invited to take online survey administered by Qualtrics^{XM} Survey Software. By opening the survey and answering a single question, participants will provide informed consent. The survey asks a series of questions grouped into categories regarding your experience with Cystic fibrosis as a male. Participation for this study is voluntary and takes about 15-30 minutes to complete online.

I will ask you questions regarding what information you received about reproductive health, reproductive options and sexual health. If you are a father of child with CF, I may ask what your family has communicated to your son regarding these topics. I also would like to know what resources you would like made available for your family by HCPs who serve you. There are no right or wrong answers. What is important are your opinions. Your responses will only be used for the purpose of this study. At the end of the survey, I will collect some personal data from you such as your age, educational background, and your/your child's CF mutation. After the interview, I may contact you to clarify certain points.

BENEFITS AND RISKS:

Most interviewees find the questions interesting. There are no risks associated with participating in this study. It is hoped that the information gained from the study will help other families with CF and their providers. There will be no cost as a result of participation in this study. In return for your time and effort you will receive a \$20 Visa Gift Card for participation in this study. If you do not complete the study, you will receive Gift Card commensurate by each quarter hour completed.

Participation in this research is completely voluntary and you may choose to withdraw from the research at any time. Participation is voluntary and you can choose not to answer questions that you do not feel comfortable.

CONFIDENTIALITY

The information you provide will be kept private and confidential. You will be given a false name and identifiable information will never be used in a publication or presentation. I will not pass on your details to any organization or company. This is a research study for academic purposes only and is not connected with the government.

CONTACT INFORMATION:

If you would like to contact me further questions about this research study, please contact me via email at dianna.sanderson@uscmed.sc.edu or by phone 561.376.2565.

If you have any questions about your rights as a research participant, you may contact a staff member of the University of South Carolina Institutional Review Board (Attn: Latoya Newton 803.576.7326 or email at (newtonla@mailbox.sc.edu).

By signing below or responding to an online question upon beginning the survey you acknowledge that you understand the explanations provided or read to you above and you voluntarily agree to participate in this study.

Signature

Date

Dianna Sanderson
Principal Investigator
Graduate Student
University of South Carolina| Genetic Counseling Program|
School Medicine

APPENDIX D: FATHER-SON RSH COMMUNICATION THEMES

Themes Pertaining to Father-Son RSH communication
These are the major themes and sub codes.

- Cystic Fibrosis
 - Body image
 - Restrictions
- Sex
 - Delayed
 - Partners
 - Monogamy
 - Multiple
 - Consensual sex vs rape
 - Dangerous sex practices
 - Safe sex
 - Healthy sex
 - Sex life
 - Sex talk
- STIs
- Esteem
 - Self esteem
 - Esteem building
- Sex Partners
 - Monogamy