

Attitudes and Decision Making Related to Pregnancy Among Young Women with Cystic Fibrosis

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Published online: 16 August 2016
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Abstract *Introduction* The number of female patients with CF able to consider pregnancy has increased with improved therapies. This study explored attitudes and decision making regarding pregnancy among young women with CF. *Methods* Twenty-two women with CF ages 18–30 years completed semi-structured, in-person interviews exploring experiences with preconception counseling and reproductive care in the CF setting. Interviews were audio-recorded, transcribed, and coded using a thematic analysis approach. *Results* Participants indicated CF is a major factor in pregnancy decision making. Although women acknowledged that CF influences attitudes toward pregnancy, many expressed confusion about how CF can affect fertility/pregnancy. Many perceived disapproval from CF providers regarding pregnancy and were dissatisfied with reproductive care in the CF setting. *Discussion* Young female patients with CF reported poor understanding of the effect of CF on fertility and pregnancy and limited preconception counseling in CF care. Improvements in female sexual and reproductive health care in CF are warranted.

Keywords Cystic fibrosis · Pregnancy · Fertility · Sexual and reproductive health

Significance

What's Known on This Subject As survival in cystic fibrosis (CF) improves with new therapies, more female patients are confronting decisions about their reproductive futures. Although the true fertility rate in females with CF is unknown, pregnancies do occur. Given the association of medical complications with pregnancy among women with CF, careful planning in this population is needed to ensure the best outcomes for mother and baby.

What This Study Adds This study investigates attitudes and experiences of young women with CF regarding pregnancy, fertility, and sexual and reproductive health (SRH) care in the current care model and explores facilitators for improvement in care.

Introduction

Advancements in therapy and management have led to dramatic increases in the life expectancy of patients with CF [Cystic Fibrosis Foundation (CFF) 2012]. If mortality rates continue to decline at the current pace, the projected median survival for patients with CF born today will be into the sixth decade of life (MacKenzie et al. 2014). In addition to longer survival, patients with CF have had progressive improvement in markers of disease severity, including mean body mass index and lung function, in the past several decades [Cystic Fibrosis Foundation (CFF) 2012].

Improved survival and overall health has led to an increased number of women with CF making sexual and reproductive health (SRH) decisions, including childbearing decisions. Little is known about CF and fertility in women (Ahmad et al. 2013). Women with CF usually have normal reproductive anatomy, but may have thicker cervical

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mucus and more anovulatory cycles than women without CF (Edenborough 2001; Johannesson et al. 1998). The impact of these mechanisms on fertility is poorly understood and the true fertility rate is unknown (Lyon and Bilton 2002). Women with CF who become pregnant have been found to have similar respiratory trends compared to non-pregnant women with CF although they may require increased care in addition to their usual CF maintenance during pregnancy (McMullen et al. 2006; Tonelli and Aitken 2007). Long-term survival does not appear to be negatively impacted by pregnancy (Goss et al. 2003). However, pregnancy and motherhood do appear to lead to more illness-related visits, pulmonary exacerbations, and a decrease in some domains of quality of life, likely reflecting the impact of motherhood on disease self-management (Schechter et al. 2013). Additionally, although relatively rare, pregnant women with CF are more likely to require mechanical ventilation, have pneumonia or acute renal failure, and die during delivery compared to women without CF (Patel et al. 2015; McArdle 2011).

With the rising number of female patients with CF confronting decisions about SRH and pregnancy, the purpose of this study was to investigate their attitudes and experiences regarding pregnancy, fertility, and SRH care in the current CF care model. This study employs a qualitative methodology to elucidate the heterogeneity of attitudes and range of preferences as well as explore patient experiences with SRH and pregnancy in the CF setting. Because there is minimal published literature about pregnancy decision making among women with CF, a qualitative study is a particularly useful approach to gain insight about this topic area and to generate hypotheses. These findings can guide future quantitative investigation around preconception care in CF.

Materials and Methods

Twenty-two women with CF aged 18–30 years were recruited to participate in individual, semi-structured in-person interviews during inpatient or outpatient visits in an accredited adult U.S. CF care center between June and December 2014. Participants were told the interviews were focused on SRH care in CF. Attitudes toward fertility/pregnancy and experiences with preconception counseling and reproductive care were explored using an interview guide with key questions and probes (Table 1). The primary investigator (PI) or a co-investigator conducted interviews after participant demographic information (including CF and SRH history) was collected. All interviews were audio-recorded and transcribed verbatim. The final sample size was guided by both a purposeful sampling strategy to ensure a heterogeneous mix of younger and older participants and

Table 1 Key interview questions

What are your thoughts on becoming pregnant?
How easy or difficult do you think it would be for you to become pregnant?
Would you like to ever become pregnant and have a child?
What factors would be important in making a decision to become pregnant? How does your CF play into your thoughts about pregnancy?
Has anyone on your CF care team ever discussed fertility/pregnancy with you?
Would you like to talk about sexual and reproductive health issues with your CF doctor?
Ideally, how would you like sexual and reproductive health care to occur? How would you want to hear about sexual and reproductive health information specific to CF?
What would you like to tell other girls or women about your experiences in discussing sexual and reproductive health with your CF care team?
What would you like to tell CF providers about your experiences in discussing sexual and reproductive health with your CF care team?
In your opinion, how can your CF doctor better support women such as yourself with sexual and reproductive health concerns?

thematic saturation (Crabtree and Miller 1999; Sandelowski 1995). Thematic saturation in this sample was reached after the thirteenth interview. The study was approved by the University of Pittsburgh Institutional Review Board (PRO14030382).

Interview transcripts were analyzed through an iterative process of coding to identify themes. Three coders (TK, BS, TG) developed an initial set of codes and then independently reviewed each transcript to apply the initial codebook and to identify additional codes. Using a consensus coding approach, the coders met to review their coding, discuss any discrepancies, and define any new codes; this was repeated using an iterative process until no new codes emerged. A senior co-investigator (EM) was available to adjudicate any differences in interpretation. The final coding scheme was applied to all transcripts and the investigative team identified central themes. Representative quotations were selected from the transcripts to illustrate themes identified. ATLAS.ti 5.0 (Scientific Software Development GmbH, Berlin) was used to facilitate data management and coding.

Results

The final sample included 22 participants, with a mean age of 25.1 ± 2.81 years (range 21–30 years). Five participants had been pregnant once and two had been pregnant twice or more. Full demographics can be seen in Table 2. Several themes surrounding pregnancy and fertility emerged from the interviews (Table 3).

Table 2 Participant demographics

Characteristic	Mean (SD) or % (n)
Age	25.1 (2.81)
<i>Self-rating of CF severity</i>	
Mild	41 % (9)
Moderate	46 % (10)
Severe	14 % (3)
<i>FEV1 (self-report)</i>	
≥70% predicted	55 % (12)
40–69% predicted	27 % (6)
≤40% predicted	18 % (4)
Age of first sexual intercourse with male partner	17.9 (2.8)
<i>Contraceptive choice(s)</i>	
Oral contraceptive pills	50 % (11)
Condoms	73 % (16)
Depo-provera	27 % (6)
Withdrawal	32 % (7)
IUD	5 % (1)
<i>History of pregnancy</i>	
Once	23 % (5)
Twice or more	9 % (2)

CF as Major Factor in Reproductive Decision Making

Participants were asked to identify factors in reproductive decision making. Most indicated that their CF disease played a major role in their thoughts and feelings about having children. When speaking about why CF is a major factor, one woman stated, “[CF] adds more gravity to the decision because it’s not just having a child, but it’s also making sure that I’m healthy enough to become pregnant.” Other factors that emerged included financial stability, maturity, and relationship status.

When asked to rank these factors, many women named CF-related health issues as the most important consideration. Specifically, participants considered potential CF complications when thinking about future pregnancies. One participant stated, “I’ve heard a lot about pregnant women with CF having a lot more complications with their health during their pregnancies. So, in a way, it makes me feel like I need to prepare myself. It adds more gravity to the decision because it’s not just having a child, but it’s also making sure that I’m healthy enough to become pregnant.” Women’s perceptions about their ability to care for children also shaped decision making. One woman described her hesitation to adopt, “Cause I’m not gonna be...say, ‘Ok, I’ll take a

Table 3 Key themes regarding reproductive attitudes and experiences in young women with CF

Theme	Quotes
CF as major factor in reproductive decision making	“I guess I’ve heard a lot about pregnant women with CF having a lot more complications with their health during their pregnancies. So, in a way, it makes me feel like I need to prepare myself. It adds more gravity to the decision because it’s not just having a child, but it’s also making sure that I’m healthy enough to become pregnant.”
Desire for a sense of “normalcy” with reproductive decision making	“They [the CF team] just kept saying—you can’t do it, you’re just gonna have to have an abortion because you’re sick, you have CF. And it was just really hard on me because I wanted to feel normal, like somebody who didn’t have CF [and] could have a baby.” “Don’t let [your patient] just assume that they’re never going to have a baby. Whether you think it’s good for their health or not, it’s [your patient’s] decision.”
Lack of communication about pregnancy and reproductive health with CF team	“I don’t think [CF providers] think about vaginas. I think they just think about lungs.” “I always thought that [CF providers] would tell me not to [become pregnant]. They’d be disapproving of it because of my health and...that my health would decline cause all those pamphlets they gave me said that women have a hard time carrying children that have CF. So, I always thought they would probably disapprove of me trying.”
Misconceptions about fertility and pregnancy leading to sexual risk behaviors	“I think—just in general addressing pregnancy and even fertility would be helpful. Just because I don’t really know much about it, regarding CF.” “Well, for a long time, I thought CF women didn’t have babies. [Then] on Facebook, I got invited to a group that was Cystic Fibrosis Moms and these women having babies have CF and...I was always under the impression that that wasn’t going to happen for me.” “I do have a daughter. We weren’t using anything at the time, because I was told I wouldn’t get pregnant without fertility [treatments] because of CF. So we didn’t worry about it, and then [I became pregnant].”
Need for sexual and reproductive health education and counseling in CF care model	“Starting the dialogue and...letting patients know that they can talk about it [sexual and reproductive health] ... I feel like I would rather my [CF] doctor make it more well-known that they have that [sexual and reproductive health] information rather than less. [T]here’s a lot of misinformation out there... I know that there was a period time that I was under the opinion that females with CF couldn’t get pregnant as easily...I think just having an open dialogue and making sure people are getting the right information is the most important thing.” “I guess that I would like more open communication. More awareness that I can talk to them [the CF team] about those things or they can help me [with] those things cause I feel like what I did—turning to the internet—isn’t always the best idea for getting factual or good information.”

baby.’ And then not be able to take care of it.” Only a few of the participants mentioned that potentially abbreviated life expectancy factored into their decisions.

Desire for a Sense of “Normalcy” with Reproductive Decision Making

While women believed that CF might make pregnancy and motherhood difficult, many participants expressed a desire for a feeling of “normalcy” with pregnancy-related decisions. One participant explained, “They [the CF team] just kept saying—you can’t do it, you’re just gonna have to have an abortion because you’re sick, you have CF. And it was just really hard on me because I wanted to feel normal, like somebody who didn’t have CF [and] could have a baby.”

The belief that women with CF have the “right to have a child” was tied to this desire for normalcy. Participants desired autonomy in pregnancy decision making and, while they appreciated the advice of family and desired input from the CF team in the context of their health, they felt that the decision to have children was theirs alone. The advice one woman gave to CF providers was, “Don’t let [your patient] just assume that they’re never going to have a baby. Whether you think it’s good for their health or not, it’s [your patient’s] decision.”

Lack of Communication About Pregnancy and Reproductive Health with CF Team

Participants reported a lack of communication with the CF team regarding pregnancy and reproductive health. Most reported a complete lack of preconception counseling and had never spoken to their CF team or any healthcare provider about their fertility or pregnancy intentions or desires. A participant who already had a child simply stated, “I don’t think [CF providers] think about vaginas. I think they just think about lungs.” One woman described never bringing up the topic of pregnancy fearing disapproval from her CF team, “I always thought that [CF providers] would tell me not to [become pregnant]. They’d be disapproving of it because of my health and...that my health would decline cause all those pamphlets they gave me said that women have a hard time carrying children that have CF. So, I always thought they would probably disapprove of me trying.” Another woman admitted not communicating with her CF provider that she had been trying to get pregnant for a year because “he was already mad about it.”

Among participants who did discuss pregnancy with their CF team, there was wide variation in how these discussions were handled. Some participants were advised against pregnancy due to poor health. In describing these discussions, some displayed emotional acceptance, while others expressed feelings of frustration, anger or sadness.

Misconceptions About Fertility and Pregnancy Leading to Sexual Risk Behaviors

Participants expressed confusion about how CF affects fertility and pregnancy. As one woman said, “I think—just in general addressing pregnancy and even fertility would be helpful. Just because I don’t really know much about it, regarding CF.” Some reported being told or believed themselves to be infertile or have low fertility due to CF. Others were unsure of their fertility, often due to conflicting information. One woman explained, “Well, for a long time, I thought CF women didn’t have babies. [Then] on Facebook, I got invited to a group that was Cystic Fibrosis Moms and these women having babies have CF and...I was always under the impression that that wasn’t going to happen for me.”

Misconceptions about fertility often led to subsequent contraceptive non-use and risk for unintended pregnancy. As one participant said, “I do have a daughter. We weren’t using anything at the time, because I was told I wouldn’t get pregnant without fertility [treatments] because of CF. So, we didn’t worry about it, and then [I became pregnant].” Several participants reported engaging in sexual risk behaviors and unprotected sex in their adolescent years due to assumed infertility and ignorance over their risk for unintended pregnancy. One woman stated, “Yeah, it [hearing about potential low fertility] hurt...hurt me a lot and then it just got to the point where I said, ‘You know what?—I was taking a chance and a risk.’”

Need for SRH Education and Counseling in CF Care Model

Given the importance of CF as a factor in pregnancy decision making, participants reported dissatisfaction with the lack of communication around fertility and pregnancy. Participants shared a clear recommendation for early education around SRH issues in CF and open, repeated discussions with the CF team. One participant described, “Starting the dialogue and...letting patients know that they can talk about it [SRH] ... I feel like I would rather my [CF] doctor make it more well-known that they have that [SRH] information rather than less...I think just having an open dialogue and making sure people are getting the right information is the most important thing.”

Many participants believed that provider counseling in this area was vital as much of the SRH information online for women with CF was incorrect. “[T]here’s a lot of misinformation out there... I know that there was a period of time that I was under the opinion that females with CF couldn’t get pregnant as easily,” one woman described. Another participant said, “I guess that I would like more open communication. More awareness that I can talk to them [the CF

team] about those things or they can help me [with] those things cause I feel like what I did—turning to the internet—isn't always the best idea for getting factual or good information.”

Discussion

Young female patients with CF report that their CF disease is a major factor in reproductive decision making. Many women desire a sense of normalcy around pregnancy and parenthood. Despite the intricate links between CF disease and reproduction, young female patients with CF have poor understanding of the effect of CF on fertility and pregnancy. Participants described limited SRH discussion and preconception counseling by their CF team. When discussions did occur, participants often perceived disapproval toward childbearing from their CF providers. Existing misconceptions around fertility and limited receipt of SRH care lead to sexual risk behaviors and unintended pregnancies in young women with CF. To address this gap in comprehensive care, young women want improved SRH educational resources coupled with routine SRH discussions initiated by their CF providers.

As this study highlights, reproductive decision making is complex for young women with chronic conditions (McNary 1999; Smeltzer 2002; Tong et al. 2015). Previous qualitative work in women with CF around pregnancy decision making in the U.K's CF care model supports our results as women with CF felt discussions around pregnancy with the CF team were crucial, but often not begun until a patient was married. Additionally, women echoed the theme of a desire for normalcy and felt that CF should not affect whether they decide to have children (Simcox et al. 2009). Prior survey-based work confirmed that parenting and fertility issues are important for patients with CF and that many participants were dissatisfied with SRH discussions with their health providers (Fair et al. 2000).

Studies have also demonstrated that women with CF may not seek evidence to aid in reproductive decision making for fear of being advised against child-bearing (Simcox et al. 2009; Conway et al. 1994). Given these perceptions, it is critical that providers convey respect for women's autonomy around pregnancy decisions. Projecting judgment or disapproval can prevent women from seeking information that can help optimize their health in the preconception period and can erode the patient-provider relationship (Stevens 2015).

This study highlights the continued gap in patient knowledge around CF and fertility/pregnancy and the effect of misconceptions on sexual behaviors. Survey work by Sawyer and colleagues 20 years ago clearly demonstrated that women with CF have poor knowledge around fertility and

pregnancy in CF (Sawyer et al. 1995). More recent work has confirmed a lack of general SRH knowledge in young women with CF (Korzeniewska et al. 2009; Gage 2012).

Preconception and general SRH care provision may be especially critical as previous literature has revealed that women with CF may often utilize less effective contraceptive methods, such as natural family planning (Plant BJ et al. 2008; Gatiss et al. 2009). A recent study found that only 49% of sexually active female patients with CF age 18–45 years were using contraception at all compared to 65% of women in the general U.S. population. In addition to contraceptive choice, nearly one-quarter of pregnancies reported in this study were terminated due to either being unintended or secondary to deteriorating maternal health status (Roe et al. 2015).

Given the complex relationship between CF disease and reproductive decision making and the possible complications during pregnancy associated with CF, routine dialogue about SRH between women with CF and their CF care teams is encouraged (Simcox et al. 2009; Frayman and Sawyer 2015; Tsang et al. 2010; Towns 2010). Current European guidelines advise that all members of the multidisciplinary CF team have a role in educating and informing the woman of the interplay between CF and pregnancy. Furthermore, preconception counseling should be offered to all patients with CF of childbearing age (Edenborough et al. 2008). The CF Foundation guidelines for adult CF care similarly advise preconception discussion of the effects of pregnancy on CF (Yankaskas et al. 2004). In light of the perceived disapproval by CF providers toward pregnancy in these interviews, it is imperative that the CF team supports pregnancy decisions in women with CF and helps patients make the best decisions to optimize their health during pregnancy.

Despite these recommendations, U.S. CF program directors identify several major barriers to SRH care and discussion in the current CF care model, including lack of adequate time during clinic appointments, discomfort of providers and patients around SRH issues, and lack of knowledge and training in SRH (Kazmerski et al. 2015). With this in mind, further quantitative investigation of provider attitudes and practices around SRH care for young women with CF is needed. Secondly, understanding patient preferences will be imperative in the development of effective interventions in the current CF care model. This evidence can guide the creation of patient educational resources and CF provider training around SRH care and discussion. Furthermore, partnerships between the CF care team and women's health specialists knowledgeable about CF (specifically maternal-fetal medicine specialists given the complexity of pregnancy in this population) should be explored as an avenue to improve comprehensive SRH care and preconception counseling.

This study has several limitations. First, the focus of qualitative research is to explore participant perspectives in

great depth; sample sizes are generally small, thereby limiting generalizability. However, these narratives are critical to understanding the complex factors that influence pregnancy attitudes, experiences, and decision making in young women with CF. Results from these interviews may not be applicable to all young women with CF as the patient interviews were conducted at a single center. Due to limited generalizability, future quantitative investigations should include a larger sample of women with CF to detect differences in SRH attitudes or preferences across relevant patient characteristics, including age, disease severity, and parity.

In the age of novel targeted medications, such as ivacaftor and lumacaftor/ivacaftor, more investigation into the physiologic effects of CF on fertility and pregnancy is justified. These agents systemically alter the basic CFTR defect and are believed to improve lung function and mucociliary clearance through effects on mucus viscosity (Rowe et al. 2014; Wainwright et al. 2015); similar improvements in mucus viscosity may occur in the cervix leading to potential effects on the fertility and SRH of women with CF (Jones and Walshaw 2015). Thus, investigation into the effects of novel personalized therapeutics on the SRH of female patients is warranted.

In conclusion, as new therapies and management lead to improved survival, women with CF are increasingly confronting decisions around their reproductive futures. CF providers are in a critical position to offer preconception counseling and general SRH care. Further investigation into the implementation of SRH care in the CF care model, including collaboration with women's health specialists, is warranted.

Acknowledgments This research was funded by the Cystic Fibrosis Foundation 1st and 2nd Year Fellowship Grant (KAZMER13B0).

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