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## Adults with Cystic Fibrosis and (In)fertility: How Has the Health Care System Responded?

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As the average life expectancy for people with cystic fibrosis (CF) continues to improve, issues of sexuality and reproduction will become increasingly relevant to this population. The majority of children with CF today will survive to adulthood and will likely face issues of sexuality and reproduction in their lifetimes. Indeed, living with CF does not preclude parenthood. Some adults with CF have children, others desire to have children in the future, whereas others do not.

Fertility issues are a strong focus among both men and women with CF. Fertility bears centrally on reproductive decision-making, determining whether natural conception is even an option or whether adoption or assisted reproductive technology must be considered. Because most adults with CF have been regular visitors in the health care system since birth or early childhood, they are certainly influenced by how that system explains reproductive health issues related to their illness. However, the extent to which health care providers have played a role in helping adults with CF understand reproductive issues such as fertility is not well understood.

This paper discusses results from an exploratory, qualitative case study of how reproduction is being addressed for adults with CF in the health care setting (Hull, 1999). The case chosen for this study was a typical CF center featuring a group of adult patients with CF and a multi-disciplinary team that included a pediatrician, pulmonology fellows, social worker, nutritionist, and nurse-practitioner. Direct observations of clinic interactions (over a 4-month period) and in-depth interviews with adults with CF (n = 10) and their various health care providers (n = 6) were conducted. Content analysis was performed on the field notes and interview transcripts. This was an exploratory study not designed to generate generalizable knowledge about adults with CF and reproduction; however, the findings highlight important themes and considerations for the provision of reproductive counseling about fertility-related issues for adults affected with CF.

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## Background

Cystic fibrosis is an autosomal recessive genetic disorder that generally affects the pulmonary and digestive systems, is most prevalent in caucasians of northern European descent, and occurs in approximately 1 out of every 2500 live births (National Institutes of Health, 1995). Although CF may be fatal in childhood, survival trends have greatly improved in recent years. The Society for Adolescent Medicine estimates that 60% of people born today with CF will survive at least to age 20 (Blum et al, 1994), and the median survival of a person with CF is 29 years.

CF affects reproductive health by presenting calculable genetic risks to offspring, delayed sexual maturation, and reduced fertility. Approximately 97% to 98% of men with CF are infertile because of abnormal development of the vas deferens, epididymis, and seminal vesicles. Azoospermia is caused by thick secretions in the vas deferens, and 50% of sperm are morphologically abnormal (Lemke, 1992). However, there are reports in the literature of men with CF with the ability to father children biologically (Barreto et al, 1991). Reproductive options for the majority of men with CF who are infertile include *not* to have children, adoption, in vitro fertilization with donor sperm, and microscopic epididymal sperm aspiration (MESA) coupled with in vitro fertilization. MESA is a relatively new surgical technique that is used to acquire viable sperm (Silber et al, 1995; Hamberger et al, 1995). Of these options, only MESA enables biological paternity for otherwise infertile men with CF.

Delayed puberty and somewhat reduced fertility are characteristic in women with CF. Although fertility rates lower than 85% for “normal,” unaffected women have been suggested for women with CF (Brugman and Taussig, 1984), the exact fertility rates and potential causes of lowered fertility are unknown (Hilman et al, 1996). Some have even suggested that fertility rates may not be significantly different among women with CF compared with the general population (Sawyer et al, 1995), although this remains controversial. Closely monitored pregnancies among women with CF are generally well-tolerated and have little effect on respiratory health status. Positive fetal and maternal outcomes are associated in particular with mild disease (Canny et al, 1991).

## Findings

Four primary themes emerged from this study regarding how CF-related fertility issues are (or are not) addressed within the health care setting, how both men and women with CF are learning about these issues, and what other information resources may be available to affected adults. These themes are illustrated with direct quotations from interviews with both affected adults and health care providers.

### Finding 1a

Conversations between adults with CF and their health care providers about reproductive issues tend to be one-time discussions that take place at the initiative of the patient rather than the provider.

**Finding 1b**

Adults with CF may not learn about important fertility issues within the health care setting unless they ask specific questions of their providers.

Discussions with patients with CF about reproduction in the health care setting are often trigger-based rather than proactive, and occur in response to a specific event or question. Providers in the CF clinic indicated that the topic of reproduction, including future reproductive potential, is often raised first in the pediatric setting with the parents of a recently diagnosed child. After this initial discussion, reproductive issues are discussed more sporadically. For example, contraception was not discussed consistently with adolescent patients. An adult pulmonary fellow in the clinic described her reactive approach to raising reproductive issues with young female patients:

I usually ask the young women about their menstrual history. And don't probe as to whether they're sexually active, specifically. But if that information were to come out, I would feel comfortable raising the issue of birth control with them, and safe sex and AIDS prevention.

There was no indication that such discussions ever occurred with young men in the CF clinic.

Some providers in the CF clinic indicated that fertility issues were raised with most patients by the time they were teenagers:

For the males, we do talk to them usually in their mid-teens about the fact that there is a very high incidence of infertility or sterility among males with CF. We also have a pamphlet that we give them. There's an introductory pamphlet on reproductive issues in CF which covers this.... We don't have, or at least I don't have any set visit or time or age at which this happens. Often it's in response to inquiries from the patient or from the family.

However, a social worker in the clinic was aware that some of their male patients did not know that they were sterile:

I've heard of some teens being surprised that, the male teens being surprised that they were sterile. And that's a really sad thing to hear. And I think that was probably an educational gap that happened with the parents, that they were uncomfortable discussing it.

Implicit in her comments is the belief, shared by other providers as well, that parents should be talking to their affected children about reproductive issues.

Some providers explained that they tend not to address reproduction in the CF clinic because of the amount of time it takes to address the medical issues facing their patients. An adult pulmonology fellow indicated, "I never thought to inquire [about reproductive issues], actually, because I get so distracted by the medical issues. And I've never heard anybody else bring it up either."

Although health care providers may not initiate conversations with their affected adult patients, they answer questions and address issues that are raised by their patients concerning reproduction. A social worker in the CF clinic expressed her concern regarding this trend:

I think it's, unfortunately, that it gets left up to, a lot of times, the adults taking the responsibility to talk about it or ask about it. I don't know how much of a priority it is for the staff, I guess.

Indeed, a physician in the clinic indicated that he addresses reproductive issues by answering his patients' questions rather than initiating conversations about reproduction:

If there is an 18, 19, 20 year old patient that we're following, particularly one who's not married, would I routinely ask, "Are you thinking about pregnancy?" No ... in those cases, I would probably be waiting for clues from the patient.

Clues may consist of direct questions from patients or knowledge of patients' romantic relationships or marital plans. In effect, this places the responsibility for initiating conversations about reproduction with the patient.

### **Finding 2a**

Men with CF may not learn about their likely infertility until they are older, sexually active, and actively planning their families.

### **Finding 2b**

Men with CF need time to adjust to their infertility and to accept the alternative reproductive options available to them.

The assumptions held by some CF providers that their adult patients understand fertility issues contradict affected adults' reported lack of understanding about fertility issues. Several men with CF in this study grew up assuming that they would be able to have biological children, learning about their likely infertility only as adults. While some seemed to take this information in stride, for others it came as a shock. A young man with CF described the unusually abrupt and insensitive manner in which he had learned about his likely infertility from his primary care physician outside of the CF clinic:

A doctor asked me if I was having sex, and I told her yes, I had been in a relationship for a while. And then she was like, "You are using a condom, right?" And I'm like, "No," because [my girlfriend] was on the pill at the time. And she was like, "Just because of your condition doesn't mean that you can sleep around with everybody." And I was like, "What does that mean?" She said, "Just because you're sterile doesn't mean you can go having sex with everybody and it's a big free-for-all." And I was like, "Whoa! What was that all about?"

Prior to this disclosure, he had never heard that most men with CF are infertile.

Receiving new information about their disease as adults, such as learning about fertility issues, could affect how an adult with CF feels about parenthood. A man with CF who had

believed that he wanted to have children in the future described what it was like to learn that he was infertile:

I had mixed results. First I was like, “Yahoo, I don’t have to use a condom or anything.” And I was being selfish: “I don’t have to worry about kids; I can keep all my money for myself.” And then I thought now, that’s not how it is. So then it struck me. I was upset. I mean, I had dealt with so much.

The realization that he was infertile caused several shifts in his conception of fatherhood and, ultimately, he arrived at a decision to pursue adoption.

Men with CF in this study talked about coming to terms with adoption or the methods to help them father a child as ways to address their infertility. Some went through a period of adjustment to learning that they are unable to father a child through more conventional means. One man with CF had not yet let go of the idea of having children through natural conception:

My doctor just let me know again that it might not be possible [to father a child through natural conception], but it may be possible to have surgery-type stuff. So that’s been in the back of my mind. But I never really took it to heart, which I still don’t.

In addition, the alternatives of adoption and MESA followed by in vitro fertilization are expensive options that require advance planning. A man with CF and his wife were exploring their options after learning from his urologist that he is infertile:

What we’ve done over this past year, we’ve looked into adoption. We started touching bases with our health care providers and seeing what they’ll cover, how much involvement they’ll take. So that’s all been within the last year. And we found that—we have some friends that are in the adoption agency, and adoption is extremely expensive .... And the biggest problem that we ran into [with MESA] is that I have one health care provider and my wife has an HMO. So they won’t—they won’t even consider working together to try infertility drugs or whatever, they won’t even consider it. That’s one of the other things that we ran into: we might be stuck for paying for one end or the other.

### **Finding 3a**

Women with CF may underestimate the degree to which they are fertile.

### **Finding 3b**

Some women with CF do not use reliable methods of birth control because they believe they are likely to be infertile.

### **Finding 3c**

Unplanned pregnancies among the women with CF may be related to unreliable use of birth control due to beliefs about CF-related infertility.

Many of the women with CF in this study described a current or former belief that it would be harder for them to become pregnant because of their illness, and some of these women indicated that they did not regularly use birth control:

When I did become sexually active, to be perfectly honest, since I had been told that it will be harder for me to get pregnant as someone with CF, getting pregnant wasn't foremost in my mind. I mean, I had friends that were taking birth control pills, and we were in high school, and I'm like, ha ha, because in my mind I'm thinking I've been told that it's darn hard for me to get pregnant. But now I look at it in a more rational and mature way, what if I was one of those people with CF who just happen to get pregnant easily?

One woman with CF explained that she did not see a gynecologist to talk about methods of contraception until she was an adult and did not use reliable methods of birth control as a sexually active teenager: "As many accidents as there have ever been in the past with contraceptive use and stuff like that, I'm surprised. I mean I'm thinking maybe I'm not very fertile." Contraceptive use had not come up during her care in the CF clinic, and it was not until she consulted a gynecologist outside of that clinic at age 18 that she had a conversation with a health care provider about birth control. Her comments also suggest a relationship between beliefs about fertility and use of birth control methods. Interactions with health care providers about contraception, sporadic as they are, seem to encourage more reliable use of birth control methods among adolescent women with CF.

A physician in the CF clinic acknowledged that about half of the pregnancies among his patients with CF were not planned:

I would say that it's a mixed bag, probably half and half I would say. At least half of them, certainly, have said, "We're thinking about getting pregnant, what do you think? Can you tell me what to expect?" This and that. And some just come in and say they're pregnant.

At least some of these unplanned pregnancies were related to a lack of contraceptive use, which was sometimes linked to beliefs about fertility.

#### **Finding 4**

Although some written educational materials are available to adults with CF about reproduction and fertility, some of these resources may be out of date or incomplete.

Written brochures, pamphlets, fact sheets, and articles were an important source of factual information about genetic inheritance and reproductive health issues for the women with CF in this study. (The men with CF in this study did not discuss such written literature as a source of information and seemed to rely more on conversations with their health care providers.) For some women with CF, written materials may be the only source of information with which they are provided, although this information is sometimes outdated or incomplete. Several women with CF remembered information on fertility and CF from older literature they had read growing up:

A long time ago in the books, they said that cystic fibrosis men can't have children or it's a very slim chance of them having children. And it's hard for a CF woman to get pregnant if she makes a lot of mucus or whatever than somebody without cystic fibrosis. But in some they don't make a lot of mucus and they could conceive a lot faster than when they don't. And I don't make a lot of mucus, so I guess I conceived.

This account of fertility for men and women with CF is fairly accurate, even though it was based on older literature. Another woman with CF, however, recalled inflated statistics on female infertility in CF: "I think I got a handout once, and I think the statistics in that were 80% of women couldn't have and 99% of men couldn't, were sterile." This suggests that one reason why women assume they are infertile may be that they have only seen older and outdated information to this effect in written pamphlets and brochures.

## Discussion

Reproductive counseling is not a priority in the specialized health care setting for adult patients with CF. Other CF-related concerns subsume the focus of care in this setting, causing important reproductive issues such as fertility to be addressed infrequently and at times that are less useful to affected adults' reproductive plans and decisions. Health care providers may raise reproductive issues at specific time points, such as at diagnosis or in the context of pregnancy; however, they often do not raise these issues when affected adults are actively making reproductive plans and decisions, except in response to specific questions. This approach to addressing reproductive issues among adults with CF may contribute to the lack of preconception planning and elements of surprise upon learning that one is (or is not) infertile.

This study highlights a concern about the need for providers to talk to men with CF about the 97% to 98% chance that they are infertile. One man with CF in this study was not aware that he was sterile until adulthood, when he learned from a primary care provider outside of the CF clinic in an abrupt and inappropriate manner. Providers in the CF clinic differed in their accounts of how many male patients are aware of infertility issues, although a social worker knew of at least some teenagers with CF who were not aware that they may be infertile. It seems this problem is not unique to the setting in which this study was conducted. The majority of men with CF and their parents who participated in another study did not know that men with CF are generally infertile (Hames et al, 1991), and others have described a widespread lack of communication between providers and male patients with CF about infertility (Sawyer, 1995).

In addition, this study underscores a need for women with CF to have more accurate information about their potential fertility. Unplanned pregnancies among the women with CF in this study were related to unreliable use of birth control due to beliefs about CF-related infertility. Many of the women with CF in this study believed that they could not get pregnant and therefore believed that they did not need to use contraception on a regular basis. Consistent with these findings, another empirical study of women with CF found that 69% believed that affected women had reduced fertility or were infertile. In addition, 33% of

the women in that study who had been pregnant did not plan their pregnancies (Sawyer et al, 1995).

Whereas Sawyer et al could not draw a direct link between beliefs about infertility, failure to use birth control, and unplanned pregnancy, the present study suggests that these 3 issues are indeed linked. Many women with CF in this study assumed that they were infertile because they had read that abnormal cervical mucus caused reduced fertility. This belief, in conjunction with a lack of information and little discussion with health care providers about contraception, led many of the women with CF in this study to use contraception in an unreliable manner.

Affected adults need accurate information about contraception, fertility, maternal risks of pregnancy (for affected women), and various reproductive options in order to appropriately plan for their family life. Women with CF who want to have children may want to pursue genetic testing of their partners prior to becoming pregnant in order to learn the risks of passing CF to their offspring. Pursuing genetic testing in the context of an unplanned pregnancy alters the range of choices available to women. Men with CF who want to have children and have an awareness of their likely infertility can pursue semen analysis. With this information, they can make choices about whether to try to have a child through natural conception, adoption, or MESA. In addition, they can also explore the various financial issues surrounding the latter 2 options. However, if affected adults are relying on outdated educational literature or if CF-related fertility issues are not consistently addressed within the health care system—as this study suggests—affected adults may not learn about these issues at a time in which the information is useful for their reproductive planning and decision-making.

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