

“If We Would Have Known”: A Couple’s Regret Over a Missed Opportunity to Have a Biological Child After Lung Transplantation

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Sigrid Ladores, PhD, RN, PNP, CNE¹,
Leigh Ann Bray, MSN, RN, CNL, PhD(c)¹,
and Janet Brown, MSN, RN, CPN¹

Abstract

Cystic fibrosis (CF) is a genetic, chronic disease that results in thickened secretions in the respiratory, gastrointestinal, and reproductive tracts. Over 95% of males with CF have congenital bilateral absence of the vas deferens causing infertility. This is a case study of a 29-year-old male who underwent a lung transplant after 8 months of oxygen dependency secondary to poor lung function. Approximately 1 year posttransplant, he and his wife decided that they wanted to start a family and consulted a fertility specialist who advised them to utilize donor sperm due to the teratogenic effects of posttransplant medications. Taken by surprise with this news, they expressed regret about the missed opportunity for pretransplant sperm aspiration and cryopreservation to conceive a biological child. He reported, “If we would have known, we would have made sure I [banked] my own [sperm].” This case study highlights a critical gap in CF comprehensive clinical care.

Keywords

cystic fibrosis, lung transplantation, fertility preservation, patient education, patient perspectives/narratives

Introduction

Cystic fibrosis (CF) is a genetic, chronic disease that affects the CF transmembrane conductance regulator protein causing abnormal ion transport across cell membranes (1). Abnormal ion transport results in thickened secretions in the respiratory, gastrointestinal, and reproductive tracts (2). Lung transplantation is a common treatment for those with severe pulmonary disease to preserve life and quality of life (3). With advancement in science, lung transplant recipients have more than an 80% survival rate at 1 year and greater than 50% survival at 9 years (4). The improvements in quality of life and survival enable recipients to consider a healthy future with a family of their own. However, posttransplant immunosuppressive medications may be teratogenic to a fetus and have been associated with preterm birth and preeclampsia (5), making timeliness of reproductive health education crucial. Therefore, patients with CF and their partners need CF-specific education regarding fertility so that they can fully explore their reproductive options prior to lung transplantation. This is a case study of a young man with CF and his wife who shared their heartbreaking journey to parenthood.

Background

Once considered a disease of childhood, more than half of the CF population is now over the age of 18 years (6). Individuals with CF want to meet developmental milestones at about the same times as their healthy peers (7). Over 95% of males with CF have congenital bilateral absence of the vas deferens causing infertility (6). With the current median life expectancy around 40 years of age (6), individuals with CF desire to experience all of life’s key events, including attending college, engaging in productive work, marrying, and planning families.

¹ School of Nursing, The University of Alabama at Birmingham, Birmingham, AL, USA

Corresponding Author:

Sigrid Ladores, School of Nursing, The University of Alabama at Birmingham, NB304, 1720 2nd Avenue South, Birmingham, AL 35294, USA.
Email: sladores@uab.edu



Case Presentation

A 29-year-old male, with CF heterozygous DF508 mutation, underwent a bilateral lung transplant in March 2013 after 8 months of oxygen dependency secondary to poor lung function. Approximately 1 year posttransplant, he and his wife decided that they wanted to start a family and consulted a fertility specialist who advised them to utilize donor sperm due to the teratogenic effects of posttransplant medications. Taken by surprise with this news, he stated: “[Having to use donor sperm] was bigger than them [fertility specialists] saying, ‘You can’t have kids.’” They expressed regret about the missed opportunity for pretransplant sperm aspiration and cryopreservation in order to conceive a biological child because of the lack of education provided by their CF care team. Although they understood that the CF care team’s main focus was on the pulmonary system to save his life, they also mourned the loss of their reproductive capability. He and his wife now want to be advocates for others who are contemplating transplantation and are in their childbearing years.

Methods

An in-person, audio-recorded interview was conducted with the husband and wife dyad who selected their own pseudonyms to ensure anonymity. The recording was transcribed verbatim and analyzed independently by the 3 authors. Inter-coder agreement was high at approximately 90%. Using Braun and Clarke’s method of thematic analysis (8), codes were developed into themes and subthemes.

Results

The overarching theme of “Needing Fertility Education Related to Lung Transplantation” emerged from the interview data, with the following 2 subthemes that describe the critical phases of their experiences: (1) before lung transplant and (2) after lung transplant.

Before Lung Transplant

Kellen (pseudonym) described that growing up with CF meant that he focused on the daily care requirements to manage CF and simply “making it day by day.” As his lung function began to deteriorate, his CF care team focused on preserving his lung function and maintaining eligibility for lung transplant. Megan (pseudonym) shared, “His family never really thought to talk to him about it [fertility], because they never thought he would make it to that.” When Megan first found out that Kellen had CF, she fearfully thought, “I’m never gonna have kids! He’s gonna die at 27!”

In fact, at age 15 when he started dating Megan, Kellen found out that males with CF are generally infertile. Kellen recalled that around age 20 when he was still dating his same high school sweetheart, his thoughts on fertility and reproduction were, “It’s way in the future, so I’m not gonna sit here and worry about it now.” Looking back on his experiences, Kellen stated, “Growing up [with CF], you feel like you can’t [have kids]. But

there’s a way, wow, there’s different options! At first you don’t realize that. I think that’s a big thing that should be talked about.”

Megan and Kellen both expressed the need for education regarding fertility options during routine CF clinic appointments. Specifically, they wished that they received counseling on fertility preservation prior to transplant so that they could have a chance to conceive a biological child. Both voiced regret about what they felt was a missed opportunity. Megan stated, “Up until you’re ready to have kids, you don’t ever see a fertility specialist. Your pulmonologist doesn’t care about that aspect of your life.” Although they acknowledged that their CF care team’s first priority was preserving Kellen’s lung function, they expressed regret about not hearing about their full reproductive options.

After Lung Transplant

Approximately 1 year after Kellen’s transplant, he and Megan made an appointment with a fertility specialist to begin planning for what they thought was still possible at that time, a biological child of their own. This was their hopeful prayer throughout their toughest days during transplant and recovery. Megan shared that, “When we got married, we were like, okay, there’s still fertility treatments. We always had this hope. Fertility [treatments]. That’s the answer.”

When the fertility specialist reviewed Kellen’s posttransplant medication regimen, he uttered the following words that shook the couple to the core: “You know what? I just don’t think it’s [using Kellen’s sperm] a good idea because it [posttransplant medication] can cause birth defects.” Kellen described that receiving this news was “the biggest blow.” He elaborated, “I almost melted in my chair and was just like ‘no!’ I literally about walked out the door.” Kellen shared his frustrations: “Your average person doesn’t even think of that. Doesn’t think what medicine would do what [to a fetus]. That’s something that we [CF care team and couple] hadn’t talked about [but] was very important.” Megan added, “I feel like as a physician, it’s your responsibility to educate us on these things [freezing his sperm], and I feel like the ball was definitely dropped.”

As they described the process of moving forward and selecting a sperm donor, Megan said, “I just wish that we could be an advocate for people knowing the effects of transplant meds before transplanting. Because if we would have known, we would have absolutely went the route to make sure Kellen could have given his own [sperm]”. She ended with, “There’s not enough education [about fertility preservation].”

Lessons Learned

In this interview, the couple shared their regret over a missed opportunity to have a biological child. Their experience revealed a need for pretransplant fertility preservation counseling and full disclosure of posttransplant reproductive options. They were unaware of the impact of immunosuppressive medications on their reproductive options post lung transplant. Having experienced the heartache of not being able to have a biological child, Kellen and Megan are now vocal advocates

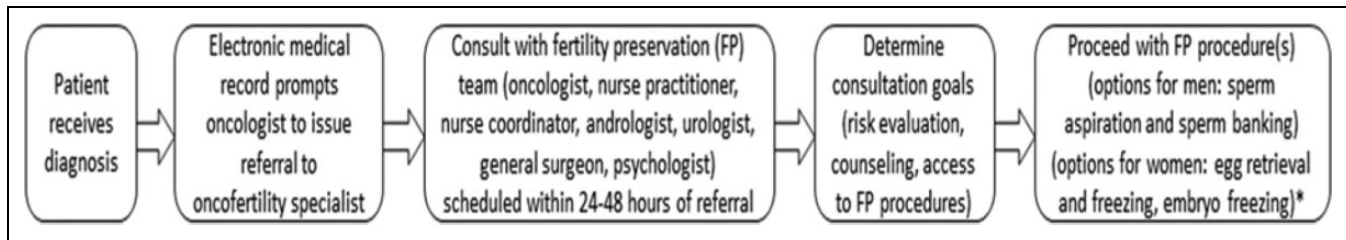


Figure 1. Fertility preservation (FP) model in oncology. Adapted from Blough et al, Colorado Oncofertility Program (10) and Carlson et al, Children's Hospital of Philadelphia's FP Service (11). *Other centers offer additional FP procedures that are currently regarded as experimental (eg, testicular and ovarian tissue cryopreservation).

for other individuals with CF and encourage them to seek comprehensive clinical care that includes discussion of reproductive health and family planning.

This case highlights how the lack of comprehensive clinical care delivery greatly changed the course of a couple's path to parenthood and affected their overall emotional well-being. The couple's narratives demonstrate the critical need for research and education in CF reproductive health issues. Their powerful story shows that the perspectives and experiences of adults with CF wishing to become parents must be elicited to inform the development of clinically relevant interventions to meet their unique needs. While fertility preservation counseling is part of standard of care for oncology patients undergoing radiation and/or chemotherapy, there are no related clinical guidelines used in patients with nonmalignant conditions (9). The fertility preservation model routinely used in oncology may serve as a template for adoption in the CF population (Figure 1).

Conclusion

With the average life expectancy of individuals with CF continuing to rise, their desire to experience all developmental milestones, including parenthood, must be considered. This case study calls attention to a crucial gap in comprehensive clinical care that requires patient-centered interventions, education, and research.

Authors' Note

The authors acknowledge that the work is their own. It has not been previously published and is not under consideration for publication elsewhere.

Declaration of Conflicting Interests

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Author Biographies

Sigrid Ladores is an assistant professor at the University of Alabama at Birmingham School of Nursing.

Leigh Ann Bray is a PhD student in Nursing.

Janet Brown is a PhD student in Nursing.