Exploring the Developmental Impact of Cystic Fibrosis on Lung Transplant Candidacy:
Considerations for Adulthood

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We have no known conflict of interest to disclose

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Abstract

The average life expectancy for cystic fibrosis (CF) has increased over the past four decades resulting in a higher rate of adult CF patients. Adults seeking lung transplant to address CF related advanced lung disease (ALD) represent a small, yet growing, subset of lung transplant recipients. Psychosocial factors such as adherence to medical recommendations, self-management of medical care, and caregiver support have been identified as positive prognostic factors in lung transplant outcomes. These factors are also implicated in the pediatric chronic illness literature and are crucial as patients begin to transition to a more autonomous and independent role in their own health management. Adults with CF facing ALD must navigate through another transitional phase as lung transplant requires additional supports and new expectations. A case series is used to highlight specific psychosocial considerations in this population and to explore the seemingly dichotomous relationship between independent self-management and caregiver support.

Keywords: Cystic Fibrosis, psychosocial evaluation, lung transplant, medical self-management, caregiver support

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Cystic fibrosis (CF) is a genetic condition which impacts approximately 35,000 individuals in the United States and is characterized by chronic respiratory, digestive, and reproductive issues (Centers for Disease Control and Prevention [CDC], 2020). CF is considered a multisystem disease involving a protein deficiency which impacts exocrine tissue (Katkin, 2020). Common associated clinical features can include sinus disease, pancreatic disease, bowel obstruction, infertility, and anemia (Katkin, 2020). Respiratory involvement is one of the hallmark associations of CF and individuals living with the condition must manage persistent and productive cough, decreased pulmonary functioning, and increased risk of airway infection (Katkin, 2020). Affected individuals are often diagnosed in infancy or early childhood (CDC, 2020). CF management involves a complex routine of nutritional monitoring, pharmacotherapy, and breathing treatments (Cystic Fibrosis Foundation [CFF], n.d.). Caregivers of young patients facilitate and oversee treatment adherence with the expectation to gradually shift responsibility to the individual as they mature. Theoretical models of pediatric chronic disease self-management have posited that patients should have more control over the management and treatment of the condition as they age and develop (Lozano & Houtrow, 2018; Henry & Schor, 2015).

The rate of patients with CF who reach adulthood has increased over the past 40 years (CFF, 2018). Recent estimates indicate that half of the current CF population is over the age of 18 (CFF, 2018). The transition from pediatric care to adult care has notable challenges for CF patients including, most commonly, a transition to new medical providers and medical home as well as greater independence and higher level of responsibility for medical care (Askew et al., 2017). A framework for CF management transition suggests that the desired transition outcome is for the patient to be an independent administrator of their own treatment with minimal

involvement from the caregiver (Leeman, Sandelowski, Havill, & Knafl, 2015; Williams, Mukhopadhyay, Dowell, & Coyle, 2007). Once the transition takes place and patients establish adult healthcare, most adults with CF are able to adjust well, exhibit adaptive coping, and engage in developmentally typical tasks (Hamlett, Murphy, Hayes, & Doershuk 1996). Despite the improvements in survival rate, CF is ultimately a fatal congenital condition and progressive respiratory insufficiency remains a major cause of mortality among patients with the diagnosis (Turcios, 2020).

Lung transplant to address advanced lung disease (ALD) is an effective, though high-risk, advanced treatment option for organ failure in CF patients to extend life (Morrell, Kiel, & Pilewski, 2019; Ramos et al., 2019). Lung transplant, as with any solid-organ transplant, is associated with risk of post-surgical infection, allograft failure, and even death (Gabardi & Tichy, 2013; Morrell et al., 2019). However, the survival outcomes for lung transplant trail behind those of other commonly transplanted organs primarily due to the heightened chronic allograft dysfunction risk in this population, which is characterized by airflow obstruction infection, and acute rejection (Todd & Palmer, 2011; Yusen et al., 2016). Potential lung transplant candidates must go through an extensive multi-disciplinary assessment to evaluate transplant candidacy prior to listing and to compare the risks of surgery against the benefits of a new lung. Potential candidates are screened for other disease processes to rule out comorbid conditions which can negatively impact transplant outcomes (Morrell et al., 2019). The pre-transplant evaluation additionally assesses for various psychosocial or psychiatric conditions which may impair or prevent the patient's ability to fully engage in the necessary recovery and management (Morrell et al., 2019; Smith et al., 2016). A clinician with expertise in the behavioral, emotional, and social aspects of transplant, such as a health psychologist, a

psychiatrist or a medical social worker, interviews the patient and their caregiver(s) to collect data regarding multiple psychosocial domains, including adherence, mental health, substance use, cognitive capacity, understanding of treatment and treatment options, ability to cope with illness, social support, and social history (Dew et al., 2018). These psychosocial domains have been previously defined and recommended in a joint collaboration between the International Society for Heart and Lung Transplantation (ISHLT), Academy of Psychosomatic Medicine (APM), American Society of Transplantation (AST), International Consortium of Circulatory Assist Clinicians (ICCAC), and Society for Transplant Social Workers (SWSW) (Dew et al., 2018).

The psychosocial evaluation is complex, and each case presents with differences in risk factors, as well as differences in severity of said risk factors. The ISHLT/APM/AST/ICCAC/STSW put together a set of psychosocial contraindications to guide decision making, the list includes non-adherence to treatment, an uncontrolled psychiatric condition directly impairing adherence, active substance use, and inadequate social support (Dew et al., 2018). There are numerous psychosocial factors associated with transplant outcomes, and these factors are often inter-related, and not always present in terms of a definitive contraindication. The evaluating clinician must weigh the information obtained from the interview to discern patient risk level.

The focus of this paper is on two specific psychosocial prognostic factors associated with improved outcomes following lung transplantation; a patient's ability to adhere to treatment and willingness to effectively engage in their health care, and the presence of a caregiver(s) to provide support and assist the patient during recovery (Barbour, Blumenthal, & Palmer, 2006; Rosenberger, Dew, DiMartini, Dabbs, & Yusen 2012). Per the

ISHLT/APM/AST/ICCAC/STSW, the role of the designated caregiver(s) is crucial and is established during the pre-listing evaluation (Dew et al., 2018). The designated caregiver(s) is expected to transport the patient to follow up clinic visits, oversee the at home treatment plan, and monitor the overall condition of the patient. Likewise, transplant candidates are expected to take an active role in their recovery and the life-long management of the new organ (Dew et al., 2018).

Adults receiving lung transplants to address CF related ALD represent a small, but growing, subset of lung transplant recipients (Benden, Goldfarb, & Stehlik, 2019). The same pre-transplant psychosocial procedures and guidelines followed for other lung pathologies, such as COPD, emphysema, and interstitial lung disease, are applied when evaluating adults with CF. However, there is an unmet need to better understand precisely how the prognostic indicators manifest and influence the likelihood of adults with CF being approved for lung transplantation or the transplant outcomes. Adherence to treatment, whether primarily managed independently by the patient or caregiver mediated, is one component of the overall evaluation process. While the psychosocial variables examined by the clinician are intricately connected, it is suggested in this paper that the role of adherence and management may be fundamentally unique in the adult CF population due to their disease course. The congenital and progressive nature of CF results in adult CF candidates presenting for transplant at younger ages than transplant candidates with other underlying lung diseases (Benden et al., 2019; Yusef et al., 2013). Younger age, in turn, is linked to higher survival rates in patients transplanted to address ALD attributed to CF (Morrell et al., 2019). Additionally, both adults with CF and their caregivers, have previous experience with complex medical treatment regimens given their history of pediatric chronic illness.

The purpose of this paper is to explore the singular pre-lung transplant psychosocial considerations associated with the caregiver and adult CF patient relationship as it relates to medical management of ALD. Specifically, we illustrate several cases that focus on the identified psychosocial prognostic factors of adherence to medical recommendations, self-management of medical care, and caregiver support. These factors will be discussed in terms of the psychosocial pre-transplant readiness evaluation and clinical implications for candidacy.

Case 1

Patient A is a 33-year-old female who was evaluated for re-transplantation to address graft failure nearly two years after her initial bilateral lung transplant. Graft failure was due, in part, to suspected poor adherence to immunosuppressant medications. Patient A had a significant psychiatric history, including a recent display of para-suicidal behaviors when she overdosed on oxycodone. Patient A's parents acted as her post-transplant caregivers. Patient A reported adequate relationships with both her mother and father, who had divorced when she was a child. Patient A described her living situation, noting that she had never lived independently, but rather would switch periodically between her parents' homes. Patient A's parents presented as a united front at the evaluation and indicated that they were willing to do everything necessary to support their daughter's medical needs. They additionally appeared hopeful that their daughter would get re-transplanted viewing it as a means for their daughter to gain more independence. Despite the consistent level of support endorsed by her caregivers, Patient A's emotional functioning at evaluation, coupled with previous graft failure, suggested possible underreporting of instabilities in her psychiatric status, self-management, and stability within the home. The pattern presented included caregiver involvement which was seemingly contingent on the acuity of Patient A's physical and emotional functioning and medical condition. In the absence of decompensation or

crisis, the degree of caregiver involvement appeared to vacillate towards under involvement. While in certain situations, such variation may be appropriate, in this case Patient A did not have the baseline self-management ability and remained more reliant on caregivers than developmentally appropriate. Decrease in caregiver support resulted in immediate decline in health. This case demonstrates that the quality of support and the adaptiveness of the caregiver role also must be considered.

Case 2

Patient B is a 35-year-old male, seeking lung transplant to address respiratory failure due to a recent CF exacerbation. Patient B had been aware of the potential need to transplant for several years prior to initiating the pre-transplant process, however he reported that he was wary about decreased independence that would come with the procedure and recovery. Patient B was estranged from his wife and living alone. He was highly adherent to his CF protocol, managing his breathing treatments three times a day. Additionally, he reported that he was able to remain adherent to his treatment regimen while working a high stress job. His mother was his identified primary post-operative caregiver and she was eager and willing to assist her son. Patient B's mother had started to clean and assist in the grocery shopping as his physical state worsened. Patient B was reluctant to allow his mother to care for him at a greater intensity level than was already occurring, despite progressive decreasing independence. Though Patient B was deemed an acceptable candidate from a psychosocial perspective, significant concerns were raised for the help rejection issues.

Case 3

Patient C is a 28-year-old male, seeking lung transplant to address a CF-related decrease in lung functioning. At evaluation, Patient C had been effectively self-managing CF and

secondary diabetes since early adulthood. Though Patient C acknowledged issues with adherence in high school, he noted that with age and maturity, he had become more accepting of his condition and was appropriately self-managing. Patient C was finishing his final year of college and had worked part-time jobs throughout his twenties. Patient C lived at home with his mother and father and was primarily supported financially by his parents due to his status as a student and his medical state, however he had long term goals of independence and was hopeful that he would achieve these goals with lung transplantation. Patient C described adaptive coping skills including spending time outside and relaxation skills. Patient C's presentation demonstrates a balance between the appropriate reliance on caregivers, while still maintaining a degree of independence and medical self-care.

Table 1

	Availability of Support/Caregiver	Receptiveness to Caregiver Support	Self-Management	Outcome
Patient A	Inconsistently available	Over reliant on caregivers (over-utilized)	Poor self-management	Over-dependent
Patient B	Available	Resistant to care (under-utilized)	Effectively self-managing	Help-rejecting
Patient C	Available	Appropriately engaged with caregiver	Effectively self-managing	Well adjusted

Discussion

In the cases described, Patients A and B depict poor balance between self-management and appropriate caregiver involvement (Table 1). Patient A had insufficient self-management and an over-reliance on her inconsistent caregivers. Patient B presented on the other end of the spectrum and was resistant to caregiver support and wary of the reduced independence that

would be required after lung transplant. Patient C integrated his own self-management and independence with support from his family in a manner that was appropriate. Moreover, Patient C recognizes the limitations and harmful consequences of being overly reliant or completely resistant to family support, and he was able to juxtapose having the ability to pursue long-term goals, while still prioritizing present day needs. In addition to the variation in patients receptiveness of caregiver support, these presented cases demonstrate the issues associated with variations in the role of the parent or caregiver. Providing support, as in the case of Patient B's mother, when unsolicited or not wanted, may create a wedge and cause the patient to reject the help, fearful of losing autonomy completely. But as described in the case of Patient A's parents, providing ineffective or misguided support can also have deleterious effects on outcomes.

The balance between caregiver support and independence must be carefully considered in lung transplant candidates. Candidates require adequate support both prior to and during the operative period, as well as beyond the acute recovery phase. Candidates must also maintain sufficient self-management skills for long term success of lung transplantation. The careful balance between these factors, support and self-management, is even more important and complex when considering the developmental phase of adults seeking lung transplant due to CF. Independence in managing one's health and treatment adherence are skills cultivated as the patient developmentally matures (McWilliams, 2019). Simultaneously, due to the course of ALD, an increase in caregiver support will be required as health deteriorates prior to transplant. Caregiver support must remain active in the acute recovery period following lung transplant. The provider conducting the psychosocial evaluation must account for these nuances when working with the patient, the transplant team, and the patient's designated support. Cases like Patient A may seem ideal from a support perspective, however upon further examination, her ability to

effectively care for herself was low, and caregiver support was inconsistent. Conversely, Patient B demonstrated adherence to treatment, but his willingness to accept assistance from caregivers was low.

Existing conceptual models are useful when considering the appropriateness of caregiver support and patient autonomy for transition out of pediatric care (Leeman et al., 2015; Williams et al., 2007). However, such frameworks do not account for the complexities of ALD or the psychosocial demands of lung transplant. Following a lung transplant referral, a patient and their primary support must adhere to a new set of medical expectations and guidelines (Weill et al., 2015). Adults seeking lung transplant to address CF related issues represent an anomalous subset of the transplant population. These are patients who are facing pre-transplant related requirements, in addition to an already time-consuming and complex treatment routine. The 2019 guidelines put forth by the CF Foundation recommend that CF specific factors be incorporated into the psychosocial evaluation, particularly assessing how living with CF has impacted adherence behaviors, psychosocial variables, and mental health factors (Ramos et al., 2019).

Implications & Future Directions

Both the average life expectancy for CF, and the number of lung transplant surgeries performed annually, have increased in recent decades (CFF, 2018; Health Resources and Services Administration, 2017). Exactly how these trends will impact number of adults with CF seeking pre-surgical evaluation in upcoming years is not yet known. A new combination medication therapy called Trikafta™, containing elexacaftor, ivacaftor, and tezacaftor, recently received FDA approval for the treatment of CF and has demonstrated positive prognostic indicators for the disease which may change lung transplant need, though it is still too early to predict (Heijerman et al., 2019). This paper echoes the recommendation of the CF Foundation

and posits the need for transplant centers to adopt a nuanced approach to the psychosocial evaluation, which is different from both the pediatric pre-transplant evaluations and from the adult pre-lung transplant evaluations to treat diseases acquired later in life (e.g. COPD). The following are recommendations for how the clinician can work within an integrated transplant service to promote the best outcomes for adult CF transplant patients.

Before the evaluation, the psychologist should work closely with the multidisciplinary transplant team in order to fully understand the patient's engagement, their health literacy, and their skill in managing their medical care. Collateral information may be obtained from the patient's social worker or transplant coordinator prior to the evaluation in order to understand their willingness to engage in medical care. In most cases, it will be crucial to consult with the patient's CF team, especially the CF team social worker, for additional data on adherence behaviors and caregiver support.

During the visit, the patient should be interviewed separately from the caregiver to assess the patient's receptiveness of caregiver support. Content related to medical management should also be elucidated to discern who is primarily making the appointments, refilling prescriptions, and communicating with the coordinator. A thorough assessment on adherence history is warranted, as non-adherence to previous CF therapies is a contraindication for listing (Dew et al., 2018; Weill et al., 2015). Factors, such as forgetfulness and minimal positive reinforcement for management efforts, are associated with non-adherence in the adult CF population (Arden, Drabble, O'Cathain, Hutchings, & Wildman, 2019; Kettler, Sawyer, Winefield, & Greville, 2002). Understanding the unique causes of non-adherence can delineate the most effective method for intervention to promote adherence and ultimately improve outcomes for the patient.

The pre- and post-transplant period is not only stressful for the patient but also for the designated caregiver, or caregivers depending on the center's support requirements. The caregivers' general understanding of the procedure and recovery should be assessed in order to cultivate realistic expectations. Depending on the caregiver plan, the clinician may need to encourage bolstering designated support to reduce the demands on the primary caregiver. The clinician can assess caregiver functioning and well-being during the visit to ensure that the caregiver has established proper self-care strategies and is utilizing adaptive coping to address the empathy fatigue and caregiver burnout that has been observed in caregivers of lung transplant recipients (Xu et al., 2012). In some cases, it may be necessary to provide additional support or refer the caregiver to their own mental health provider. The clinician can provide brief guidance and education to the caregivers to promote a caregiving style that is consistent, offering a reliable level of care in a structured environment, while remaining flexible enough to account for the normal fluctuations in functioning during the pre- and post-transplant process.

Finally, in addition to educating the patient and family about the emotional challenges associated with lung transplant, the clinician can educate the attending physicians and the transplant team about the different standards and expectations in this subset of lung transplant candidates. As the patient's physical condition declines and subsequently improves following transplant, all members of the transplant team can encourage the greatest level of patient independence that the medical state will allow. Designated clinicians including psychologists, psychiatrists, and social workers should inform other healthcare providers of the appropriate roles and balance between caregiving support and independent self-management as the patient prepares for and recovers from transplant.

Conclusion

Lung transplant candidates are a heterogeneous group with various disease processes contributing to ALD. Adults with CF related ALD comprise a subset of the pre-lung transplant candidates. The pre-lung transplant psychosocial evaluation is an opportunity for the transplant psychologist to screen for potential emotional and behavioral risk factors associated with negative outcomes. Adherence and social support are two evidence-based prognostic domains associated with transplant outcomes that are explored in this case series as they relate to developmental phase and severity of chronic illness. Adults with CF, and their designated support persons, must navigate through a new set of management expectations and treatment recommendations as they face ALD. The cases described in this paper were selected to present self-management and caregiver-initiated management as equally necessary facets of both CF and transplant care that should co-occur harmoniously. This paper suggests that the degree of self-management and caregiver-initiated management should fluctuate depending on CF status and maturity of the individual, and that when there is a discrepancy between the two (as in the case of Patients A and B) outcomes can be impacted negatively.

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