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Position paper: Models of post-transplant care for individuals with cystic fibrosis



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ABSTRACT

There is no consensus on the best model of care for individuals with CF to manage the non-pulmonary complications that persist after lung transplant. The CF Foundation virtually convened a group of international experts in CF and lung-transplant care. The committee reviewed literature and shared the post-lung transplant model of care practiced by their programs. The committee then developed a survey that was distributed internationally to both the clinical and individual with CF/family audiences to determine the strengths, weaknesses, and preferences for various models of transplant care. Discussion generated two models to accomplish optimal CF care after transplant. The first model incorporates the CF team into care and proposes delineation of responsibilities for the CF and transplant teams. This model is reliant on outstanding communication between the teams, while leveraging the expertise of the CF team for management of the non-pulmonary manifestations of CF. The transplant team manages all aspects of the transplant, including pulmonary concerns and management of immunosuppression. The second model consolidates care in one center and may be more practical for transplant programs that have expertise managing CF and have access to CF multidisciplinary care team members (e.g., located in the same institution). The best model for each program is influenced by several factors and model selection needs to be decided between the transplant and the CF center and may vary from center to center. In either model, CF lung transplant recipients require a clear delineation of the roles and responsibilities of their providers and mechanisms for effective communication.

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1. Background

Cystic fibrosis (CF) is a genetic disease caused by mutations in the cystic fibrosis transmembrane conductance regulator (CFTR) gene leading to multisystem manifestations. Most people with CF experience progressive lung disease and die from respiratory failure or undergo lung transplant. However, as a multisystem disorder CF also affects the liver, gastrointestinal (GI) tract, sweat glands, sinuses, reproductive system, and pancreas. As adult CF lung transplant recipients experience a median survival of 10 years, these extra-pulmonary manifestations of CF are increasingly important to manage [1]. Specifically, nutritional considerations (e.g., weight management, vitamin replacement), GI concerns (e.g., distal intestinal obstructive syndrome, gastroesophageal reflux, gastroparesis), sinus disease, CF-related diabetes, bone health, and mental health warrant specialized attention [2]. How to best integrate and manage the non-pulmonary elements of CF care following lung transplantation is a topic of international concern.

There is no consensus on the best model of care for individuals with CF to manage the non-pulmonary complications that persist after lung transplant. Survey data (Supplement text and Tables S4 and S4) show that most clinicians would find shared care of CF lung transplant recipients to be helpful [3]. The most appropriate model for people with CF will depend on many factors including resources, knowledge, and experience of the transplant center, the CF center, and the individual needs and circumstances of the person with CF. Any care model should be determined through shared decision making between the transplant recipient, family, and CF and transplant care teams. The purpose of this position paper is to review models of care that may be used by CF and transplant centers that share or coordinate care of lung transplant recipients with CF to provide comprehensive care for this multisystem disease.

1.1. Scope of this document

This document describes potential models of care for individuals with CF who have received a lung transplant and is limited to how health services might be delivered between the CF center and the transplant center.

This document does not address the clinical management of CF individuals after transplant nor how these models might be implemented, nor any role for primary care physicians. Guidelines for care of the CF lung transplant recipient [2] and recent experience with and guidelines on the use of elexacaftor/tezacaftor/ivacaftor [5–10] have been published and are beyond the scope of this position paper. Guidelines for CF post-transplant model of care implementation will be required but is beyond the scope of this position paper.

2. Methods

In June 2020, the CF Foundation virtually convened a group of international experts in CF and lung-transplant care. A preliminary literature search was conducted by the CF Foundation prior to this meeting to investigate shared models of post-transplant care. The committee shared the post-lung transplant model of care practiced by their programs, including the strengths and weaknesses of these models. The committee then developed a survey that was distributed internationally to both the clinical and individual with CF/family audience, to determine the strengths, weaknesses, and preferences for various models of post-lung transplant care. A summary of the results of this survey are found in the supplemental materials. Based on the results, and committee discussion of the strengths and weaknesses of distinct care models, the committee outlined two main models of post-transplant care: i) Fully integrated shared care model between transplant centers and CF cen-

ter and ii) transplant team manages and coordinates all aspects of transplant and CF care.

The committee outlined the pros and cons of both models, how the model might work in ideal circumstances, and the staffing and/or expertise that would be needed to ensure optimal care within each model. This position paper was distributed for public comment on February 9, 2022. The public comment draft was distributed to international reviewers through the CF Foundation listservs, Community Voice, the ECFS listservs, and the Cystic Fibrosis Medical Association. The committee reviewed and acknowledged and/or addressed each of the comments received during the public comment.

3. Results: models of care for CF lung transplant recipients

The essential components of multi-disciplinary CF care after lung transplant should be established to ensure that all aspects of this multi-system disease are optimally addressed. Regardless of models delineated in Fig. 1, clear communication channels between the CF and transplant care teams should be established. Table S1 summarizes the care needs and expertise required for care of lung transplant recipients with CF. Some of these guidelines were developed for pre-transplant CF individuals and application of these consensus statements to post-transplant individuals may not be directly applicable but can be used as a guide. Table 1 outlines some variables for consideration in the identification of the optimal care model for an individual with CF.

Based on the post-transplant care requirements in Table S1, the following models of post-transplant care were discussed:

- i Fully integrated shared care model between transplant center and referring CF center
- ii Transplant team manages and coordinates all aspects of transplant and CF care.

The strengths and weaknesses of these different models of care are outlined below and shown in Table 2. Regardless of the model of care chosen, there was consensus that:

- i Lung transplant team should manage the pulmonary complications of lung transplantation indefinitely.
- ii Timely and clear communication between the transplant team and the referring CF center is essential.

Both models of care have strengths and weaknesses, each is a viable option, and the choice should be individualized based on factors listed in Table 1. The best model for each patient should be chosen by the CF center and the transplant center and may vary from center to center. For some centers, a hybrid model with components of model 1 & 2 may be the best option. Irrespective of which model is chosen, providers will need to consider interpretation of pulmonary function tests among sites (different pulmonary labs and home studies in particular) with attention to potential bias and need to compare studies based on location. More importantly, each patient should have a named CF or transplant physician who is the lead coordinator in their post-transplant management. Registry monitoring of long-term transplant outcomes will be important to determine which model is best. Centers should have a mechanism to monitor patient feedback on their model.

3.1. Model 1: fully integrated shared care model between transplant center and referring CF center

In this model of care, CF transplant recipients will attend their transplant center for all transplant-related issues, predominantly pulmonary complications and complications related to immunosuppression. For the non-transplant related conditions and the extra-pulmonary complications of CF, recipients will continue to

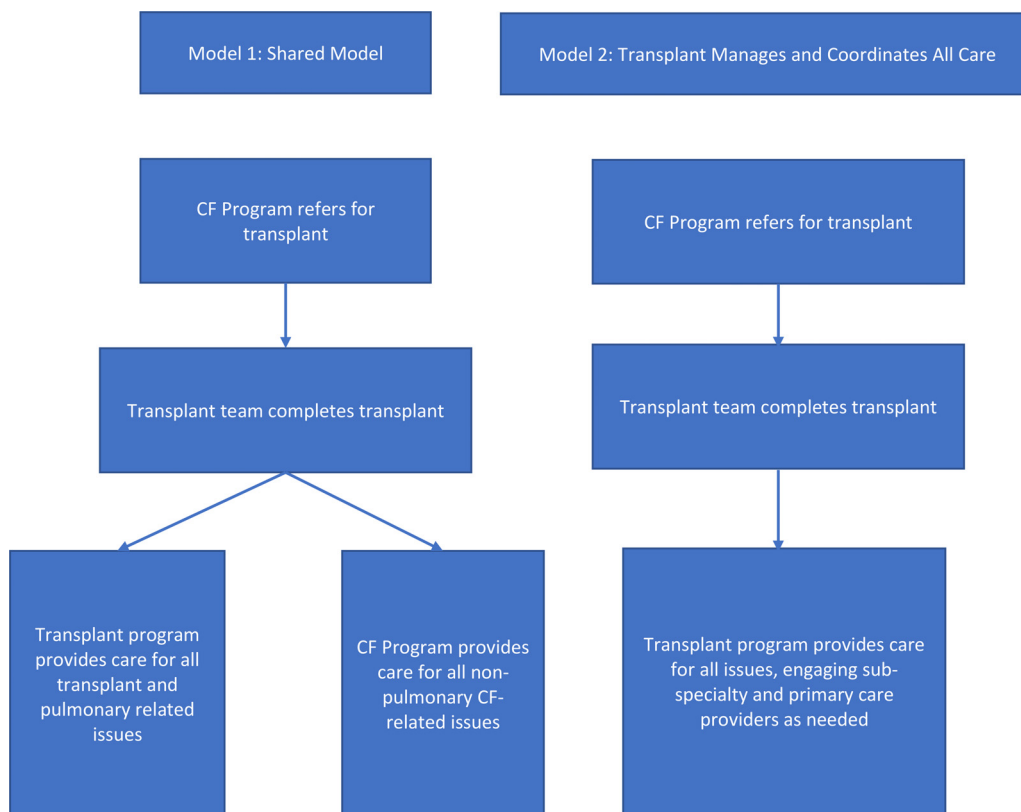


Fig. 1. Models of care after lung transplant for the individual with CF.

Table 1
Factors influencing the selection of a post-transplant CF care model.

Factor	Implications for post-transplant CF care model
Longitudinal care approach at transplant center Knowledge of CF and transplant management	Designated longitudinal outpatient care provider with expertise in CF versus group practice model for routine outpatient longitudinal care may affect quality of communication between CF and transplant centers Transplant team knowledge of CF and CF team knowledge of transplant medicine are important factors in the selection of the model of choice. Training and additions to staff may be necessary for each team depending on the model selected.
Geography	Distance to Lung Transplant and CF Centers may influence practicality of follow-up at one versus both centers
Insurance	Varies by region. Mainly an issue in United States. May limit options for follow-up locations and choice of care providers
Local health care system	Access to tertiary care hospital, primary care providers and appropriate specialists depends on local health care system (related to geography, above)
Financial resources	Out of pocket expenses for costs of care, travel, lodging near Transplant or CF center and availability of resources from fundraising efforts may impact feasibility of either model
Patient preference Physician preference	Patient's trust and acceptance of their post-transplant model of CF care Physician's commitment to post-transplant model of CF care and interest/ability to provide longitudinal care
Access to subspecialists with transplant experience (for example, nephrology, infectious disease, endocrinology, dermatology)	Local subspecialty access and expertise will vary from one institution to another such that choice of subspecialist will require careful consideration and communication between CF and transplant providers. While aspects of routine subspecialty care may be managed well at locations other than transplant institutions, providers should be cognizant that in general, transplant institutions have access to subspecialists with knowledge and experience with transplant recipients that is often not present at other institutions.
Social Support	Childcare needs, employee-employer relationship when taking time off from work, and other social factors may influence feasibility of each model at the individual patient level (related to geography and financial resources, above)

be managed by their CF center, either at the referring center or another CF care center (Table 3).

Effective communication between the CF and transplant teams is essential and, optimally, should be established prior to transplant. Intentional, formal communication between the teams should be continued indefinitely after transplant for routine elements of care, such as social work, endocrinology, GI, and other subspecialty care. In each center, a single named physician or transplant coordinator should be identified who will coordinate the care of each patient after transplant.

Pre-transplant: The CF team, in conjunction with the transplant team, helps with preparing the individual with CF and their family for identification, prevention, and treatment of the following common complications that can occur around the time of transplant:

- Distal intestinal obstruction syndrome (DIOS)
- Gastroparesis
- Diabetes: identification, monitoring, and management of hyper/hypoglycemia
- Sinus disease

Table 2
Strengths and weaknesses of different models of care.

	Model 1: Shared care model between transplant centers and referring CF center	Model 2: Transplant team manages and coordinates all aspects of transplant and CF care.
Common strengths	<ul style="list-style-type: none"> • Expert care received for both transplant and non-transplant related complication of CF • Continues existing patients/family and provider relationship and facilitates establishment of long term relationships with care team members • Execution of either model increases confidence for individuals with CF, family and health care providers that all needs can be met 	
Pros of one model over the other	<ul style="list-style-type: none"> • Facilitation of shared knowledge between transplant and CF teams • Facilitation of communication between the individual with CF, family and teams • For individuals with CF living a distance from transplant centers, reduced travel for patient with CF-related complications • For individuals with CF living a distance from transplant centers, ability to be near family and friends during transplant-related hospitalizations • Continuity of care in CF center for non-pulmonary complications of CF such as diabetes, sinus disease and GI/liver disease • Extensive education about CF and Lung transplant complications and treatment (including use of CFTR modulator) is not required for all members of both teams • Lung transplant team knows how and when to contact CF team and vice versa 	<ul style="list-style-type: none"> • Continuity of care by single transplant multi-disciplinary team • Convenient for individuals with CF/family • Potentially reduces cost to individual with CF • Facilitates communication and reduced clinic visits as occurs at single site • Efficient for medical records
Cons	<ul style="list-style-type: none"> • More than one team managing the patient, with the potential for poor communication leading to management errors • Need to attend more than one hospital with different physicians involved in different hospitals • Difficulties with overlap between transplant and non- transplant related complications of cystic fibrosis 	<ul style="list-style-type: none"> • Limited number of CF transplant providers to make this a universally viable model • Requires engaged local provider if transplant recipient lives a distance from transplant center • Potential inexperience of transplant center in managing non- transplant related complications of CF, including the use of CFTR modulators, especially if not co-located with CF center.
How this model will work in ideal circumstances	<ul style="list-style-type: none"> • Ideally, occurs in an institution with both CF and transplant teams having established, effective communication between transplant and CF multi-disciplinary team • Effective communication is paramount and best modes and patterns of communication are established • Consideration of telehealth/regular case conferences • Access to clinical records at both sites 	<ul style="list-style-type: none"> • Transplant Center and CF Center in same institution with shared positions with expertise in Transplant and CF (e.g. Dietitians, Mental health providers) • Clear communication is paramount, especially for patient geographically distant that may receive aspects of care in local CF center or with local non-CF pulmonologist • Consideration of telehealth/regular case conferences • CF specific CME with education/training for transplant team

• Mental health concerns

Immediate post-transplant period: To avoid overwhelming the individual with CF and family during the demanding early post-transplant period, the CF team should be engaged only if acute CF-related issues requiring prompt attention develop (e.g. DIOS).

Post-transplant: Informal communication must occur between transplant and CF teams immediately after transplant. At one to three months after transplant, the CF team must receive a complete and formal summary from the lung transplant team about the transplant, current issues, complications, and main medical aspects that require longitudinal follow-up (including decisions related to use of CFTR modulators after transplant). Ideally, a post-transplant CF clinic appointment would occur in parallel and on the same day (if in the same institution) with the post-lung transplant schedule within 6 months following transplant.

Beyond the first 1 to 2 years after transplant, this could be operationalized as quarterly visits, split between the CF and transplant teams, with telehealth as an option for stable patients. In-

dividuals with CF may also need to attend their local CF center prior to 6-months after transplant or visit their specialty provider with CF experience for input from non-pulmonary specialists such as endocrinology, GI, and ENT. For CF individuals attending non-pulmonary specialists at their CF center prior to transplant, continued follow-up after transplant, if needed, in their CF center institution has advantages, unless specific transplant-related complication expertise is only available at the transplant center. New non-pulmonary specialist referral after transplant should be coordinated between the transplant and CF center to ensure care is delivered in a way that promotes good communication between the non-pulmonary specialist and the transplant and CF centers. In the perioperative period, or for acute situations, while the patient remains at the transplant center, local expertise should be involved.

Geographic considerations: One of the benefits of this model is the improved management of CF transplant recipients who live a distance from their transplant center, which may constrain routine travel to the transplant center. While not ideal, for these patients, closer follow-up in the referring CF center may be the only option,

Table 3
Proposed roles and responsibilities under Model 1: Fully integrated share care for management of CF lung transplant recipient.

Team	Aspects	Medication adjustments and prescriptions
Lung Transplant	Pulmonary Decline in spirometry Symptoms Radiographic changes (e.g. chest x-ray, CT chest) Transplant Immunosuppression management (e.g. therapeutic drug monitoring) Rejection concerns (e.g. bronchoscopy with transbronchial biopsies; treatment decisions) Safety monitoring with labs (e.g. creatinine, complete blood count, liver enzymes) *Malignancy screening Infection *Pulmonary *Sinus Transplant-related (e.g. CMV) *Mental health concerns *Social work needs	• Immunosuppression Treatment for rejection Prophylactic/treatment antibiotics
Cystic Fibrosis	Endocrinology: Diabetes Bone Health Gastroenterology/Hepatology: Nutrition (e.g. weight, vitamin levels) Gastroparesis DIOS *Colorectal cancer screening Liver Disease *Mental health concerns *Social work needs *Sinus disease *(In)fertility/pregnancy/fathering concerns	Insulin and other diabetes management • Treatment for bone disease • GI medications, including pancreatic enzyme replacement therapy Vitamin supplements Chronic sinus management

*May be shared between CF and transplant teams, as outlined by local expectations for responsibilities.

even for transplant-related complications. In this setting, the transplant center, after the acute period post-transplant has elapsed, reviews the patient routinely with the CF team and sees the recipient less frequently. For transplant-related complications occurring between transplant visits, the individual with CF could attend their local CF center and receive their transplant-related complication management from the CF team, in close liaison with the transplant center. This would require a named CF physician in the CF center with interest and experience in transplant medicine including procedures and skills outlined in Table S2. This requires close communication between the CF physician and transplant team, as recommended in this fully integrated shared model of CF care. Transplant-related complications that could not be managed in the local CF center would result in prompt transfer of the patient to a transplant center. In the case of geographic barriers to routine transplant center follow-up, it may be of benefit for the transplant team to provide the CF team with up-to-date education on current practice in transplant medicine. Management of transplant-related complications in a CF center will only occur if the CF physician is adequately trained in the management of transplant-related complications. If this is not the case, the individual with CF should travel to the transplant center for management of all transplant-related complications.

3.2. Model 2: transplant team manages and coordinates all aspects of transplant and CF care

In this model, all components of CF multi-disciplinary care are located at the transplant center. While limited to a few transplant centers, a requirement is that the primary longitudinal physician overseeing CF lung transplant recipients and the transplant multi-disciplinary team has expertise in both lung transplant and CF such that they are competent to address all aspects of routine post-transplant and CF-related care and make referrals to appropriate consultants when needed (Table S1). Ideally, lung recipients for

this model live near the transplant center and would have been referred for transplant by a nearby CF program or a CF program at the same institution, in some cases with shared overlapping providers and staff with expertise in both CF and lung transplant. In institutions with CF and transplant centers in the same hospital, there are often multi-disciplinary care team members who know the transplant recipient from their pre-transplant care and can provide longitudinal input that spans the pre- and post-transplant time periods. A good example of this is a CF dietician who has time allocated to help manage post-transplant nutritional needs.

For transplant centers not in the same institution as the referring CF center, the transplant physician should have expertise in CF management and have the resources required to fully care for CF Lung transplant recipients including a CF/transplant multi-disciplinary team and access to non-pulmonary specialty physicians as listed in Table S1. Non-pulmonary specialist input that was based at the CF center should move to the transplant center to ensure all aspects of post-transplant care are centralized.

Communication between the CF team and post-transplant teams, optimally established prior to transplant, can be continued after transplant for consultation on other elements of post-transplant care, such as social work, endocrinology, and other subspecialty care, even if the transplant team is managing all aspects of CF care. A single named physician should be identified to coordinate the care for each transplant recipient. It may be useful to engage a CF-experienced pharmacist if the transplant team continues, resumes or initiates CFTR modulator therapy after transplant.

3.3. Other considerations in shared models of care

3.3.1. Communication

In 2016, the US CF Foundation launched the CF Lung Transplant Initiative (CFLTI) which aims to maximize the opportunity for transplant as a life-sustaining therapy, extend post-transplant survival, and improve quality of life for individuals with CF. To inform

the direction of the CFLTI, the CF Foundation created and funded a Consortium of academic transplant centers dedicated to improving lung transplantation outcomes and establishing a robust clinical research infrastructure. Based largely on feedback from individuals with CF and their families, a primary focus of the CF Lung Transplant Consortium (CFLTC) Quality Improvement (QI) efforts is to improve the transplant journey, particularly, the experience of transition from CF center care to transplant center care and subsequent shared post-transplant care. To support this aim and foster a culture of effective, ongoing QI at CFLTC sites, the CF Lung Transplant Transition Learning and Leadership Collaborative (LTT LLC) and Regional Dissemination Network (RDN) were established. Through the CF LTT LLCs and RDN, transplant centers work in partnership with referring CF Care Centers to improve communication, education, and relationships between CF and Lung Transplant Care Teams. This QI network has expedited the implementation of recently published CF and transplant-focused clinical guidelines and sharing of best practices for treatment of CF advanced lung disease and lung transplantation.

This work organically evolved to focus not just on the initial transition through the transplant referral process, but shared care between CF centers and transplant centers after transplant. Several key themes persist in optimizing the lung transplant journey from referral through post-transplant management including intentional communication between the CF and transplant teams; establishment of relationships between CF and transplant care providers; development and exchange of discipline-specific education tools between CF and transplant teams; inclusion of individuals with CF and their support persons in CF and transplant QI teams.

Best practices identified through the QI work include:

- Regularly scheduled virtual meetings between CF and transplant programs (quarterly or monthly)
- Virtual monthly educational teaching sessions for CF and transplant teams - each month focuses on a different topic and is presented by an 'expert' in the field from one of the centers (CFRD, GI/Nutrition, Chronic rhinosinusitis, optimizing medications in the post-transplant CF patient, etc.)
- "Refer back" form or a post-transplant handoff sheet
- Create and provide contact list of transplant care team members to CF care team and vice versa
- Co-management of CF transplant recipient document - describes what tests and support are required after transplant and who is responsible

3.3.2. Expertise and continuing medical education

Maintaining knowledge of CF and transplant medicine is critical to the success of both models especially model 2. CME is available for CF and lung transplant providers and teams interested in learning more about special considerations for caring for individuals with CF who undergo lung transplantation. The most comprehensive information is available through the Division of Continuing Medical Education at the Indiana University (IU) School of Medicine, with sponsorship from the Cystic Fibrosis Foundation and in partnership with IU eLearning and Design Services (<https://medicine.iu.edu/cme/specialized/cystic-fibrosis>). These CME courses are intended to support CF team physicians and interprofessional team members in their efforts to provide the most up to date care for individuals with cystic fibrosis. In addition to formal CME programs, the CF Foundation also convened a multidisciplinary working group that developed guidelines on post-transplant care for individuals with cystic fibrosis [2]. The North American Cystic Fibrosis Conference and European Cystic Fibrosis Society annual meetings also provide excellent education for multi-disciplinary CF Care.

3.3.3. Pediatric considerations

Since it is common for the pediatric transplant center and CF center to be geographically distanced, ongoing, intentional communication between the CF center and the transplant team is of the utmost importance. While local CF teams can contribute to the evaluation of individuals with CF at the time of an acute illness, support ongoing pulmonary surveillance (e.g., pulmonary function tests), obtain respiratory viral samples when indicated, and manage other ongoing complications of CF (e.g., endocrine, GI), it remains critical to coordinate management with the transplant team. Since there are no clear transplant-specific recommendations for nutrition in pediatric CF transplant recipients, the individual with CF's local CF dietitian can provide ongoing guidance.

In age and developmentally appropriate adolescents transition to adult transplant care may occur in tandem with transition to adult CF care. However, because of the small number of pediatric lung transplant centers, it is not uncommon that adolescents, still followed by pediatric CF centers, are transplanted at adult transplant programs. Published data suggest that outcomes for children, both on the waitlist and after lung transplant, are better when they are transplanted at a pediatric transplant program and when possible, that should be the first choice [4]. When it is not possible it is vital that pediatric CF programs work diligently with the adolescent and their family to prepare them for the transition to an adult transplant program eventually as the care after transplant will be shared between the two programs. It is equally incumbent on the adult transplant programs to include pediatric care providers on their team to provide the team with pediatric specific expertise.

3.3.4. Preferences of individuals with CF

The logistics behind Individuals with CF receiving efficient and effective care can be extremely complex. While a health care provider may structure their program(s) based on available resources, patients do not typically have the luxury of choosing many options and may be limited predominantly by two factors: geography and insurance.

From an Individual with CF's perspective there are advantages and disadvantages to each model.

With respect to Model 1, the fully integrated shared care model, where the transplant center manages all post-transplant care with a CF team managing non-transplant related CF manifestations, the advantage is that the patient will have the expertise of both trusted CF-specific and transplant practitioners. If the CF team and the transplant teams are distinct, a major advantage is having input from specialized care teams with extensive experience in lung transplant or CF. While this model allows for members of the CF team that are more familiar with the transplant recipient to continue care, the time and travel burden may be considerable.

Another challenge to Model 1, the fully integrated shared care model, is communication for various needs. This model requires delineation of first contact for transplant or CF-related concerns, which may not obviously fall to the transplant or CF program. CF and transplant concerns are often interrelated. Transplant recipients are best served by having a plan for communication based on the preferences of the transplant and CF programs.

In Model 2, where the patient has one CF Transplant team, the main advantage is obvious: ease of care and having one point person oversee all issues and concerns. Also, this facilitates building core relationships, which means not having to provide the same information to multiple providers. With respect to cost, insurance may dictate who can provide care; individuals with CF who have private health insurance are compelled to use only those programs that are covered so having care at one institution is simpler for insurance coverage.

Crucial to either care model is the relationship and trust that CF transplant recipients develop with each individual team mem-

ber. Whether that consists of one cohesive team at one hospital with one point of contact, two separate teams at two different hospitals, or a combination of the two, communication between all team members must strive to be as open and streamlined as possible and that must also include the recipient, whose responsibility is to communicate concerns clearly and concisely and to consider the opinion and direction of all team members to make the best possible decisions and to ensure the most efficient and effective care.

4. Conclusions

Care of CF manifestations in lung transplant recipients should follow best practices in the management of the non-pulmonary aspects of CF. Two models are proposed to accomplish optimal CF care after transplant. The first model incorporates the CF team into the care of CF lung transplant recipients and proposes delineation of responsibilities for the CF and transplant teams. Model 1 is reliant on outstanding communication between the teams, while leveraging the expertise of the CF team for management of the non-pulmonary manifestations of CF. The transplant team manages all aspects of the transplant, including pulmonary concerns and management of immunosuppression. The second model may be more practical for transplant programs that have expertise managing CF and have access to CF multidisciplinary care team members (e.g., located in the same institution). The best model for each program is influenced by several factors and model selection needs to be decided between the transplant center and the referring CF center and may vary from center to center. In either model, CF lung transplant recipients require a clear delineation of the roles and responsibilities of their providers and mechanisms for effective communication.

Declaration of Competing Interest

Authors declare that they have no conflict of interest.

CRedit authorship contribution statement

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Supplementary materials

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