

Development of a decision aid for adult cystic fibrosis patients considering referral for lung transplantation

Context—Most adults with cystic fibrosis are eventually required to make a decision about referral for lung transplantation.

Objective—To identify the decisional needs of these patients and to develop a decision aid to address these needs.

Methods—A comprehensive review of the literature, a review of Canadian transplant statistics from 2002 to 2006, and a self-assessment survey of patients who had already made a decision about referral were performed to identify the decisional needs of patients. A decision aid was then developed and evaluated by an expert panel of health care professionals and patients.

Results—Transplant referral patterns vary widely among Canadian cystic fibrosis clinics. Canadian patients with cystic fibrosis who were not residing in transplant centers between 2002 and 2006 were significantly less likely to undergo lung transplants ($P < .001$). Decisional needs identified by patients included wanting more information on (1) relocation to the transplant center, (2) the benefits and risks of surgery, and (3) how to cope with anxiety and depression when making the decision. In response to these identified needs, a decision aid for lung transplantation was developed. A panel of health care professionals and patients reviewed the decision aid and agreed that the content was appropriate, easy to understand, and unbiased.

Conclusion—The decisional needs of patients with cystic fibrosis who are considering lung transplantation are not being addressed in Canadian cystic fibrosis clinics, especially in clinics outside of transplant centers. An evidence-based decision aid could serve as a useful tool to help address these needs. (*Progress in Transplantation*. 2010;20:81-87)

Katherine L. Vandemheen, BScN, Shawn D. Aaron, MD, Charles Poirier, MD, Elizabeth Tullis, MD, Annette O'Connor, PhD

The Ottawa Health Research Institute, University of Ottawa, Ontario (KLV, SDA, AO), Centre Hospitalier de l'Université de Montreal, Quebec (CP), St Michael's Hospital, Toronto, Ontario (ET)

Corresponding author: Katherine Vandemheen, BScN, The Ottawa Hospital, General Campus, 501 Smyth Road, Ottawa, Ontario, Canada K1H 8L6 (e-mail: kvandemheen@ohri.ca)

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Patients with cystic fibrosis ultimately succumb to chronic airway infection, inflammation, and respiratory failure in early adulthood. Lung transplantation can be the only option left once all other medical therapies have been exhausted. For the appropriate candidate, lung transplantation can improve pulmonary function, exercise tolerance, quality of life, and survival,¹ but lung transplantation is fraught with hazards. The risk of infection and rejection are ever-present, and survival beyond 5 years is realized by only half of recipients.²

In Canada, 5 transplant centers perform bilateral lung transplantation on patients with cystic fibrosis. Because Canada is a geographically large country, many patients being referred for transplantation must move to the transplant city while they await their surgery. Furthermore, even after transplantation, they must remain in the transplant city for 3 to 6 months for post-operative transplant care. This move can have a huge emotional and psychological impact on the patients'

families and can pose a large financial burden. Consequently, the decision to be referred and ultimately have bilateral lung transplantation is very difficult for most patients and their families and they may require decision support.

Recent years have seen the emergence of shared decision making, a process whereby decisions are shared by patients and clinicians, informed by the best evidence available, and weighted in light of patients' individual characteristics and values. Evidence suggests that the quality of the decision-making process can be improved by the use of decision aids.^{3,4} Decision aids are interventions designed to help people make specific and deliberative choices among options by providing them (at minimum) information on the options and outcomes relevant to the person's health status.⁵

Little is known about the decisional needs of individuals being referred for lung transplantation. To our knowledge, no decision aids or evidence-based tools

are available to assist with this difficult decision-making process. The main objective of this study was to identify the decision needs of patients with cystic fibrosis being referred for bilateral lung transplantation and to develop a decision aid to meet these decisional needs.

Methods

The decisional needs of patients with cystic fibrosis who are considering referral for lung transplantation were identified by using a comprehensive review of the literature, a review (Canadian Cystic Fibrosis Foundation, unpublished data, June 2008) of Canadian transplant statistics from 2002 to 2006 and a self-assessment survey of patients from the Ottawa cystic fibrosis clinic who had already made a decision about referral. The study was approved by the research ethics boards of the participating hospitals and all patients who participated in the study signed written, informed consent.

An English language search of the Cochrane Library, CINAHL–Cumulative Index to Nursing and Allied Health Literature (1982-2008), and MEDLINE (1966-2008) databases was performed. Search terms used included “decisional support needs,” “decision aids,” “decision support tools,” and “decision making.” These terms were combined and exploded with the terms “cystic fibrosis,” “lung transplantation,” and “chronic lung disease.” The World Wide Web was searched by using the key words “decision making,” “decision tools,” and “lung transplantation,” and hand searches of journal articles were also conducted.

The total number of lung transplants performed from 2002 to 2006 in Canada for patients with cystic fibrosis was obtained from the Canadian Cystic Fibrosis Foundation. Further information was obtained from the individual transplant centers to identify the place of residence from which the transplant recipient was referred. This information was mapped and distance from the transplant center was calculated to determine which patients lived within a 2-hour drive from the transplant center.

Three patients who had already made a decision about referral were interviewed individually by one of the researchers. The interview consisted of questions that required both specific and open-ended responses about their perceptions of making a decision and the resources available to assist with the decision. The interviewer recorded the answers to the questions verbatim.

A steering committee composed of a nurse, 2 academic cystic fibrosis physicians/researchers, a lung transplant surgeon, and an expert researcher in decision making oversaw all aspects of development of the decision aid. The development of the cystic fibrosis decision aid followed the general format that is used for many of the other aids developed in Ottawa and was based on the Ottawa Decision Support Framework⁶⁻⁸ and guided by the International Patient

Decision Aid Standards as shown in the Table.^{9,10} The goal of the decision aid was to address the baseline decisional needs identified by the patients and the literature review by providing information on the options and possible consequences of referral and lung transplantation.

As a first step, the steering committee met and reviewed the literature and the information that had been received from clinical colleagues and the cystic fibrosis patients to determine what content would be included in the decision aid. A draft decision aid was prepared by the members of the steering committee and then reviewed in a face-to-face meeting by the whole committee to further assess its content, readability, and ease of use.

The next step was to have an expert independent panel review the draft decision aid. The expert panel consisted of 4 cystic fibrosis health care providers (2 physicians, 2 cystic fibrosis nurse coordinators) from across Canada and 4 cystic fibrosis patients who had already made a choice regarding referral for lung transplantation (1 patient had declined referral for lung transplantation, 1 patient had just returned from the initial assessment of referral, and 2 patients had received lung transplantations). The expert panel was asked to evaluate the decision aid for appropriateness, ease of understanding, acceptability of the content, and decision support methods contained in the decision aid. To elicit their opinions, they were asked to complete standardized questionnaires.^{11,12} All feedback elicited from the independent expert panel was analyzed and reviewed by the steering committee and their comments and suggestions were incorporated as design changes to produce the final decision aid.

The final decision aid was then translated to French. This translation was further reviewed and revised by 2 French Canadian cystic fibrosis clinicians. An additional version, which presented alternative survival statistics, was created for patients chronically infected with *Burkholderia cepacia* because survival of these patients after lung transplantation differs from the general cystic fibrosis population.¹³

Results

The literature review yielded 1 study that explored informational needs of cystic fibrosis patients making the lung transplant decision. Moloney and colleagues¹⁴ studied 22 cystic fibrosis patients who were transplant recipients or were awaiting lung transplant. Most transplant candidates and recipients stated that information on practical issues such as sources of financial assistance and relocation to the transplant center were integral to making the transplant decision, yet these issues were inadequately addressed by the local cystic fibrosis clinic and by the transplant program. The authors of the study concluded that, “the decision-making process

Table Methods for developing decision aid according to the International Patient Decision Aid Standards

Standard (treatment)	Method
Is based on user's needs	Patient survey: 3 patients who had made the decision at an adult cystic fibrosis clinic by using a semistructured interview guide Record review: Number of patient referrals and number of transplants linking to primary care cystic fibrosis center
Uses up-to-date scientific evidence	Comprehensive review
Has balanced, understandable content that supports decision making 1. Describes condition and natural course, decision, options, procedures involved, positive and negative features 2. Presents probabilities as event rates using a common denominator, time period, scale 3. Uses balanced presentation (easy to compare options; equal detail for positive and negative features) 4. Helps people to think about the positive and negative features that matter most 5. Includes scientific references, date of last update, readability levels, developers' credentials and conflicts of interest	Ottawa template used 1. Describes cystic fibrosis and the process of being referred vs not being referred; the pros of being referred and having successful lung transplantation include living longer and having healthy lungs; the cons of being referred and having lung transplantation include the need to move closer to the transplant center while awaiting transplant, surgical complications, and the possibility of organ rejection 2. Probabilities presented as event rates during a 10-year period by using words and pictorial diagrams 3. Options compared side by side in equal detail 4. Patients asked to rate the personal importance of benefits and risks 5. Includes references, date of publication (2006), and developer's credentials
Is reviewed by people who previously faced the decision	Survey: 4 adult patients with cystic fibrosis and 4 cystic fibrosis specialists reviewed the decision aid and completed the acceptability questionnaire
Field testing shows the decision aid is acceptable to users	
Evidence shows that the decision aid leads to informed, values-based decisions	In progress, the decision aid is being evaluated in a randomized controlled trial

could be facilitated and enhanced by practices that accommodate diverse informational needs in terms of content, quantity, timing and modality.” Evidence from other lung transplant studies suggests that patients and family members require that information be provided well in advance of the decision as they need time to think about their future and how their life will change.^{15,16}

The cystic fibrosis patients interviewed in our study about their decisional needs had the following comments:

Moving to the transplant city was very disruptive to my family life. I was unsure what to do and how we were going to manage financially.

I was lacking information about what others decided as the group [meaning the patients with cystic fibrosis] is not that big and I didn't know others who have had to make the decision. I would have preferred to talk to someone who had already been through the process.

Another suggested that making the decision about referral “might lead to anxiety and depression.”

All of the patients stated that very little information was available about the benefits and risks of the

surgery and that they would have liked to have information materials available that would have helped them better prepare when making the decision to be referred or not to be referred. They identified booklets, pamphlets, and resources on the Internet as potential information materials that could be developed.

We then set out to identify regional variations in access and referral to lung transplantation across Canada. In 2006, 3782 Canadian patients were being treated in Canadian cystic fibrosis clinics. Of these, 1547 patients (41%) received their care from a cystic fibrosis center located more than 2 hours from a lung transplant center. However, review of Canadian lung transplant statistics revealed that only 45 of 186 cystic fibrosis patients (24%), who received a lung transplant in Canada from 2002 to 2006 came from a cystic fibrosis center located more than 2 hours from a lung transplant center. Canadian patients with cystic fibrosis who were not residing in transplant centers between 2002 and 2006 were significantly less likely to have lung transplants ($P < .001$) than were their counterparts who were residing within 2 hours of a lung transplant center.

Description of Decision Aid

The decision aid was developed as a paper-based booklet that is interactive, self-administered,

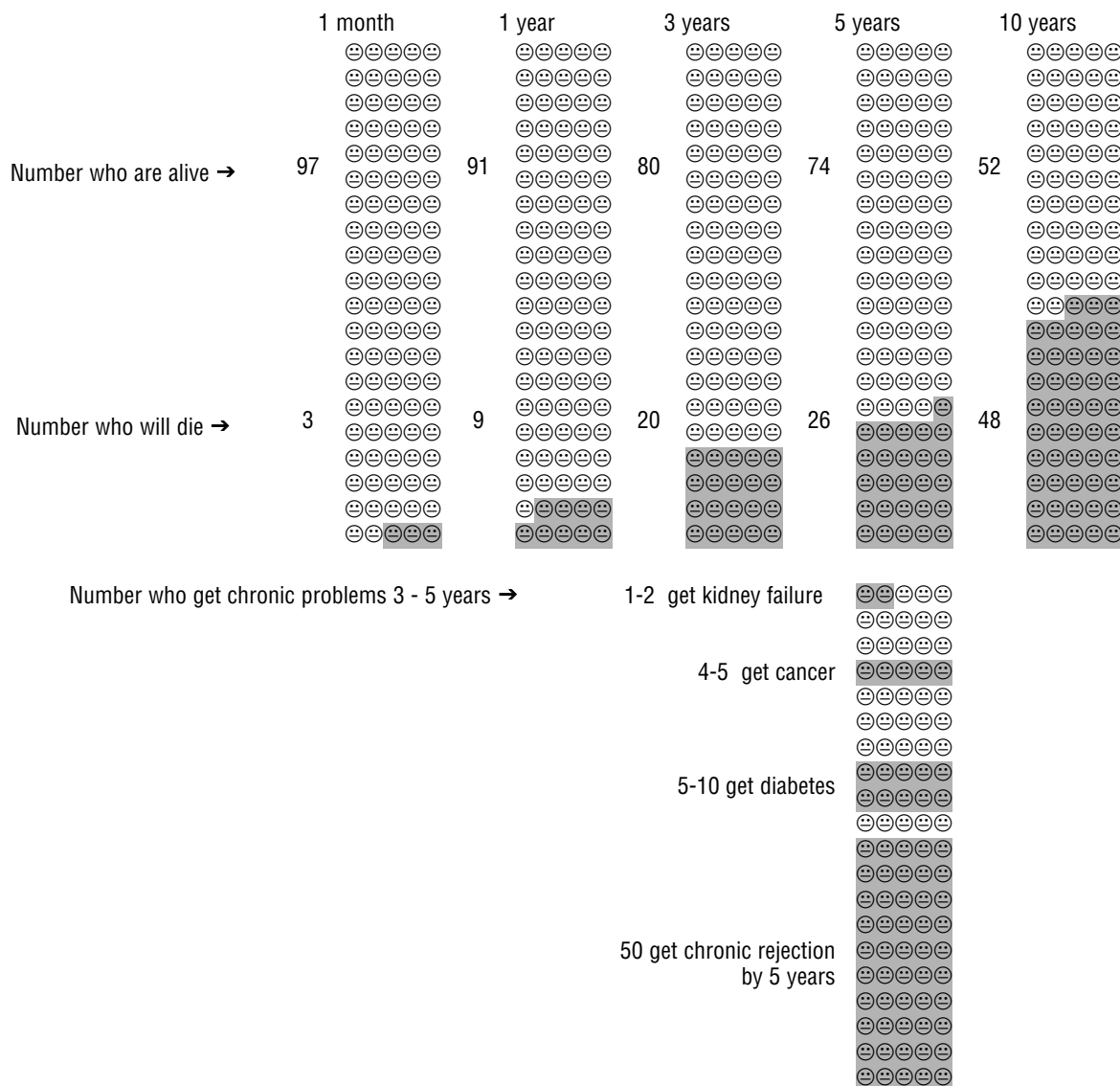


Figure 1 Expected results after lung transplantation as presented in the decision aid. Toronto Lung Transplant Program survival statistics at 1 month, 1 year, 3 years, 5 years, and 10 years for cystic fibrosis patients with no colonization of *Burkholderia cepacia* having bilateral lung transplantation in 2006.¹⁸ The lower section shows the number of patients who experience chronic problems between 3 and 5 years after transplantation.

and self-paced. A Web version is also available at <http://decisionaid.ohri.ca/decaids.html>. The paper-based booklet was chosen for its portability and low relative costs for reproduction. The Web-based version was chosen to encourage use of the decision aid by young persons who are computer savvy. The decision aid asks the patient to work through 5 steps when considering the options of “Not to be referred for lung transplantation” or “To be referred for lung transplantation.” In step 1, patients are asked to think about how cystic fibrosis affects them now. In step 2, they are asked to think about the options, benefits, and risks of lung transplantation. The pros and cons of each option are presented by using illustrative icons for the survival statistics and long-term complications in Figure 1.

Step 3 of the decision aid asks patients to choose the role they prefer in decision making. In step 4, they are asked to find out what else they need to prepare for decision making, and in step 5, they are asked to plan the next steps once they have made their decision.

Questionnaire Results

The expert panel of 4 health care professionals and 4 patients reviewed the paper-based decision aid and responded to the standardized questionnaire to evaluate the decision aid for appropriateness, ease of understanding, and acceptability of the content and decision support methods.

The 4 patients on the expert panel rated the information in the decision aid as having “about the right

information.” Three rated the length of the decision aid as “just about right” and the other respondent thought it should have “been a bit longer.”

Two of the 4 patients thought that the information provided was “completely balanced,” and 1 felt the information was a “little slanted towards referral and lung transplantation” with the fourth stating that it was “clearly slanted towards referral and lung transplantation.”

Three patients responded that “everything was clear” in the information provided in the materials, and the fourth patient stated that “most things were clear.” All of the patients thought that the information material was “very helpful” for making a decision about the options and that they would “definitely recommend” the material to other people facing the same decision.

The health care expert panel had similar responses, with 2 people rating the decision aid as having “about the right information” and the other 2 rating the information as “a little less than needed,” indicating that more information was needed on postoperative complications and long-term complications.

Three experts on the panel rated the length of the decision aid as “just about right” and the other thought it was “a little too long.” Three health care providers thought that the information provided was “completely balanced,” and 1 indicated that the information was a “little slanted toward referral and lung transplantation.” All 4 health care providers indicated that the decision aid presented the “options in a neutral and balanced manner.” All members of the health care panel thought that the information material was “very helpful” for patients making a decision about the options and that they would “definitely recommend” the material to other patients facing the same decision.

On the basis of the results from the expert panel, changes were made to the decision aid: additional information about postoperative complications was included and a pictorial presentation that included risks of long-term complications of diabetes, cancer, and kidney failure was added to the survival statistics page.

To address the concern that the information provided in the decision aid was “a little slanted towards referral and lung transplantation,” the format of the presentation was changed so that information about the benefits and risks of choosing not to be referred for lung transplantation preceded the information on benefits and risk of choosing to be referred for lung transplantation (Figure 2). The terminology used in the decision aid was reviewed again to ensure that it was consistent throughout the document.

Discussion

This study was intended to identify the decisional needs of patients with cystic fibrosis being referred for bilateral lung transplant. Once we identified specific

needs, we developed a decision aid to meet those needs. Before development of our decision aid, few decision support tools were available to these patients.

Our study showed huge variation in referral patterns across Canada. More than 75% of cystic fibrosis patients who received lung transplants from 2002 to 2006 were referred from a cystic fibrosis clinic near the transplant center. Those patients who live far from a lung transplant center may not be informed of the option for lung transplantation, or they may be more likely to choose to decline referral because of an imbalanced or biased presentation of the benefits and risks of transplant from their local center. It is hoped that a decision aid that is available to all patients on the Internet and presents information in an unbiased manner would help inform patients with cystic fibrosis who are considering lung transplantation to make a decision.

Limitations

Our study had potential limitations. The literature on decision aids recommends that a formal needs assessment and focus groups be performed to identify specific needs of the group in question. Because of the small number of patients with cystic fibrosis and advanced lung disease who are close to being considered for transplant, we were unable to perform the needs assessment completely on many patients. However we felt strongly that by interviewing 7 subjects with advanced lung disease who had already come to a decision, and by having an expert team of cystic fibrosis researchers and clinicians develop the tool, that we could identify the major decisional needs to be addressed in a decision aid. Because of the high rate of transmission of bacterial endobronchial infections between patients in this population, we were unable to hold patient focus groups.

Further Research

Our next step will be to formally assess whether this decision aid would be a helpful adjunct in the decision making process. To do so, we will conduct a randomized controlled trial of the decision aid versus standard care. All patients in Canadian and Australian cystic fibrosis clinics whose lung function (measured as the forced expiratory volume in 1 second) is less than 40% of predicted and who have not made a decision about referral for lung transplantation will be invited to participate. Each consenting participant will receive a lung transplant education and counseling session with their cystic fibrosis physician and team immediately before randomization. After the session, they will be randomized to receive the decision aid or “usual care” information. Those randomized to the “decision aid” will receive a paper-based version and a link to a secure Web site that contains the interactive decision aid. Those randomized to “usual care” will receive an information pamphlet from the Canadian

1. Not to be referred for lung transplantation		
<ul style="list-style-type: none"> ◦ You will continue to receive the same care that you have now. ◦ You need to understand that if lung function has fallen to less than 30% of normal, then 50 in 100 patients will die within 2-3 years and 50 in 100 will be alive. ◦ You will continue with your usual day-to-day activities (work, school) as long as possible. ◦ As your shortness of breath gets worse, you may need more aggressive and frequent treatment with oxygen, antibiotics, and chest physiotherapy and you may require more frequent hospitalization. ◦ Eventually, your breathing will become more labored. At this point, to help ease your shortness of breath, you will be treated with oxygen and/or a face mask breathing machine (BiPAP). If you have pain or severe shortness of breath, you will be treated with medications to help ease the discomfort. ◦ The goal is not to cure, but to provide comfort and maintain the highest possible quality of life for as long as possible. 		
2. To be referred for lung transplantation		
First assessment with the transplant team	Average time is 7-10 days	<ul style="list-style-type: none"> ◦ You go to a transplant center in Vancouver, Edmonton, Toronto, Winnipeg, or Montreal to see if you are eligible for lung transplant. ◦ You have tests of the lung, heart, kidney, and liver. ◦ You see the transplant team. You may see the social worker, psychologist, and psychiatrist to assess whether you and your family have the financial and emotional support to cope with the stress of the transplant. ◦ At the completion of the assessment, the transplant team discusses your test results with you and your family. ◦ If you are eligible but not sick enough, you will return home and the transplant team will monitor your health every 3 to 6 months until they think you should go on the transplant list.
Being put on the transplant list	Average time on the transplant waiting list is 6-18 months	<ul style="list-style-type: none"> ◦ When you are eligible and sick enough, you are put on the lung transplant list. You will need to carry a pager or cell phone 24 hours a day, and you and your family will need to live within 2 hours of the transplant center while waiting for your new lungs. ◦ Unfortunately, some people die while waiting for a lung transplant.
Lung transplant surgery	Average time in surgery is 6-8 hours Average stay in intensive care unit after surgery is 1-4 days Average time in hospital after surgery is 2-4 weeks	<ul style="list-style-type: none"> ◦ Your new lungs will come from a person who has recently died whose family has agreed to donate their lungs for transplant. You will require a general anaesthetic for the surgery. Your diseased lungs will be removed through a large chest incision. ◦ You will wake up in the intensive care unit with a breathing tube in your windpipe and you will be on a mechanical ventilator (machine that helps you breathe) for 1 to 3 days. You will have tubes in your chest (chest tubes) and catheters in your arms (intravenous) and wrist (arterial).
After hospital	Average time is 3-6 months	<ul style="list-style-type: none"> ◦ You will have to live in or very near your transplant center for several months after your transplant.
<p>After successful lung transplantation You will no longer need to do chest physiotherapy, take nebulized antibiotics, or use supplemental oxygen. You will be required to take multiple pills (at least 6 types) for the rest of your life to help reduce infection and reduce the risk of your body rejecting your new lungs.</p> <p>Although your lungs will be healthier, you will still have cystic fibrosis. Lung transplantation will not fix other health problems common in people with cystic fibrosis such as diabetes, digestive problems, osteoporosis, or male infertility.</p>		

Figure 2 Benefits and risks of lung transplantation as presented in the decision aid.

Cystic Fibrosis Foundation that describes lung transplantation. All participants will complete standardized questionnaires before randomization and 3 weeks after randomization to determine whether use of the decision aid increases knowledge about the options, improves realistic expectations, and decreases decisional conflict in comparison to usual care.

Conclusions

Despite the study's limitations, the results of this study are important. We assessed regional disparities in referral for lung transplantation and identified decisional needs of the cystic fibrosis community. The development of our decision aid is the first step forward toward providing a shared decision-making

evidence-based tool to a vulnerable population of patients who are making what is arguably the most important decision of their lives.

Acknowledgments

The authors thank the cystic fibrosis patients and the cystic fibrosis specialists who provided invaluable assistance with the development of the decision aid: Neil Brown, MD, University of Alberta; Pearce Wilcox, MD, University of British Columbia; Nancy Morrison, MD, Dalhousie University; Patrice Kean, BScN, London Health Science Centre; and Josette Salgado, RN, University of Alberta.

Financial Disclosures

This study was funded by The Ontario Thoracic Society and The Physicians' Services Incorporated Foundation.

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