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Development and Evaluation of a Decision Aid for Cystic Fibrosis Patients Considering Referral of Lung Transplantation

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Development and Evaluation of a Decision Aid for Cystic Fibrosis Patients Considering Referral for Lung Transplantation

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Abstract

Objectives: 1. Identify the decisional needs of cystic fibrosis patients considering referral for bilateral lung transplantation and develop a decision aid to meet these needs. 2. Evaluate decision aid in a prospective trial.

Conceptual Framework: Ottawa Decision Support Framework

Setting: Canadian and Australian Cystic Fibrosis clinics

Methods: Phase 1. Comprehensive literature review, Canadian transplant statistics review and a survey identified the decisional needs and a decision aid was developed.

Phase 2. The decision aid was evaluated in a randomized trial involving 149 patients.

Results: Phase 1. A wide variation in transplant referral patterns was uncovered; patients not residing in transplant centers were significantly less likely to undergo lung transplants ($P < 0.0001$). Phase 2. Patients randomized to the decision aid had greater knowledge ($P < 0.0001$) and more realistic expectations ($P < 0.0001$) compared to those randomized to usual care. Decisional conflict score was significantly lower in the decision aid group 3 weeks post randomization (11.6 vs 20.4, $P = 0.0007$) and the decisions were durable 12 months after randomization ($P = 0.06$).

Conclusion: Transplant referrals vary widely. Patients receiving usual care have unmet decisional needs. The decision aid enhanced informed decision making when combined with usual education and counseling. Future research is warranted to evaluate its implementation in clinical care.

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To my husband **Rob**, and our son **Dain**, all I can say is it would take another thesis to express my deep love for you. Your patience, love and encouragement have upheld me, particularly in those many days in which I spent more time with my computer than with you.

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List of Abbreviations

APN	Advanced Practice Nurse
CF	Cystic Fibrosis
CI	Confidence Interval
CNA	Canadian Nurses' Association
CFTR	Cystic Fibrosis Transmembrane Conductance Regulator
FEV_1	Forced Expiratory Volume in one Second
FVC	Forced Vital Capacity
IPDAS	International Patient Decision Aid Standards
ODSF	Ottawa Decision Support Framework
RNAO	Registered Nurses Association of Ontario
SD	Standard Deviation

Chapter One

Overview and Organization

Chapter 1 presents an overview of the thesis and a description of the organization of the thesis elements.

Lung transplantation for advanced lung disease in patients with cystic fibrosis (CF) has become relatively common with over forty-five lung transplants being performed annually in Canadian CF patients (Canadian Cystic Fibrosis Foundation, 2007) and over two hundred being performed in all of North America (The International Society for Heart and Lung Transplantation, 2009).

Currently, lung transplantations are performed with considerable success and many patients have an improved quality of life (Lanuza, Lefaiver, & Farcas, 2000); however, transplantation will always remain a perilous and frightening experience for patients and their loved ones (Ullrich, Jansch, Schmidt, Struber, & Niedermeyer, 2005).

Previous studies of CF patients and providers (Moloney, Cicutto, Hutcheon, & Singer, 2007) have identified that patients with CF experience considerable anxiety and uncertainty related to making decisions concerning lung transplantation. In 2004, the CF team at the Ottawa Hospital expressed concern that CF patients who were excellent candidates for lung transplantation were refusing to go to the lung transplant center for a referral consultation. Although the CF team respected the patient's decision to refuse referral, their challenge was to fully explore the patients decision making needs to ensure that they were

providing information about the risks and benefits of lung transplantation in an unbiased manner.

There is very little published information on decisional needs of patients with CF who are considering whether or not to be referred for a transplantation consultation. Therefore, effective tools are needed to help patients and their families make what may be the most important decision of their lives. Decision aids have been shown to help patients to understand choices, outcomes and probabilities and to clarify which positive and negative features of options matter most too them (O'Connor et al, 2009). However, none have been developed for patients considering lung transplantation (A-Z Inventory of Decision Aids, 2009).

The objectives of this thesis were to: 1) identify the decisional needs of patients considering referral for bilateral lung transplantation for endstage CF and to develop a decision aid to address patients' decisional needs; and 2) evaluate the decision aid in a prospective trial. The Ottawa Decision Support Framework (ODSF) was chosen as the conceptual framework to guide all phases of the project (O'Connor et al., 1998). The research conducted to accomplish these objectives is summarized in the following chapters.

In Chapter 2 the reader is introduced to the disease of CF, the state of lung transplantation in patients with CF and the decision process in lung transplantation. An overview of decision aids and the Ottawa Decision Support Framework is presented.

Chapter 3 includes the paper which has been accepted for publication in the Journal Progress in Transplantation submitted May 4, 2009. This paper,

"Development of a decision aid for adult cystic fibrosis patients considering referral for lung transplantation" addresses the first objective of this thesis by identifying the decisional needs of patients considering referral for lung transplantation and then moving forward to develop a decision aid to meet the information needs. It outlines the methods used to develop the decision aid to address the decisional needs identified by patients with CF who had already made a decision about referral for lung transplantation and those identified by the healthcare professionals who work with patients with CF. The paper also outlines the information that is included in the decision aid as described by the International Patient Decision Aid Standards (IPDAS) (Elwyn et al, 2006). A copy of the lung transplant decision aid is included in Appendix A and the IPDAS Checklist is included in Appendix B.

Chapter 4, is the evaluation paper, which has been accepted for publication by the American Journal of Respiratory and Critical Care Medicine submitted June 5, 2009. This paper, "Randomized trial of a decision aid for cystic fibrosis patients considering lung transplantation" addresses the second objective of the thesis evaluating the decision aid in a prospective randomized controlled trial. The purpose of this trial was to determine whether use of the decision aid improved the quality of decision making of 149 CF patients when considering referral for lung transplantation, in Canada and Australia compared to usual care. The primary objectives were knowledge about patient options, realistic expectations, and decisional conflict. The secondary objectives included

patients' stated choice among the options, and durability of the decision at twelve months.

Chapter 5 provides a general discussion that integrates the findings from the study with implications for nursing, practice, education, policy and research.

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Chapter Two

Research Problem and Conceptual Framework

Chapter 2 presents an overview of the problem and describes the conceptual framework guiding the research.

Cystic Fibrosis

Cystic fibrosis is the most frequently inherited fatal disorder among Caucasians (Boat, 1997). It is an autosomal recessive multi-system disease characterized by a defect in the CF transmembrane conductance regulator (CFTR) gene; a chloride-transporting channel (Davis, Drumm, & Konstan, 1996). This chloride-transporting channel is situated on the apical membrane of bronchial epithelial cells and normally functions to increase the chloride and water content of respiratory secretions (Groman, Meyer, Wilmott, & Zeitlin, 2002; Stutts, Canessa, & Olsen, 1995). The defect in the CFTR leads to increased viscosity and retention of respiratory secretions and eventual chronic infection and inflammation within the entire bronchial tree (Matsui, Grubb, & Tarran, 1998; Smith, Travis, Greenberg, & Welsh, 1996).

In Canada, it its estimated that one in every 2,500 children are born with CF. Approximately 3,400 children, adolescents, and adults with CF attend 38 specialized CF clinics in Canada (Corey, 1999).

Over the past forty years, the median age of survival has improved from fourteen years in 1969 to age thirty-seven in 2007 in Canada and the United States (Cystic Fibrosis Foundation, 2004). With this improved survival, this disease has been transformed from a disease with very high infant mortality to a

disease that must now be considered a progressive and chronic lung disease. However eventually most patients with CF develop end-stage bronchiectasis and obstructive lung disease that cannot be improved with conventional medical therapy (Allen & Visner, 2007). When conventional medical regimens fail and disease progresses in patients with severe lung disease, lung transplantation is the only option left.

Lung Transplantation

In the appropriate candidate, bilateral lung transplantation can improve pulmonary function, exercise tolerance, quality of life, and survival (Nicod, 2006; Tantisira, Systrom, & Ginnus, 2002). Nonetheless, lung transplantation remains an imperfect technique. The risk of infection and rejection are ever present (Estenne & Kotloff, 2006) and despite the advances seen in heart and liver transplantation in the last decade, no such improvement has been seen in lung transplantation survival (Liou, Adler, & Huang, 2005). Overall survival after lung transplantation internationally has remained around 50% at five years post-procedure (International Society for Heart and Lung Transplantation, 2007). Of these survivors 50-60% suffer from bronchiolitis obliterans syndrome leaving them with potentially significant morbidity (Nicod, 2006).

Recent studies have suggested potential decreased survival with lung transplantation in children with CF but no randomized controlled trial has been done to assess lung transplantation versus usual care (Liou, Adler, & Huang, 2005; Liou, Cahill, Adler, & Marshall, 2002). Given the current acceptance of the procedure nationally and internationally, a trial is unlikely to be conducted.

The Decision Process in Lung Transplantation

Decision making is the process of choosing between alternative courses of action (including inaction) (O'Connor, Jacobsen, & Stacey, 2002). Little is known about the decision making process of choosing to refer a patient for lung transplantation. Current recommendations regarding the timing of listing for transplantation is described in published guidelines from the International Society for Heart and Lung Transplantation (Maurer, Frost, Estenne, Higenbottam, & Glanville, 1998) and from the American College of Chest Physicians (Yankaskas, Mallory, and the Consensus Committee, 1998). These guidelines suggest selection criteria based on survival as well as relative and absolute contraindications (Liou, Cahill, Adler, & Marshall, 2002) (Appendix C). But the guidelines do not address the fundamental question of how patients come to the decision to choose an aggressive intervention versus pursuing palliative care for their end-stage lung disease. Since lung transplantation carries significant inherent risks, patients who are considering referral and lung transplantation must weigh their current state of disease trajectory and quality of life with potential gains of increased strength, energy, independence and ability to breathe without supplemental oxygen (Limbos, Joyce, Chan, & Kesten, 2000; Smeritschinig et al. 2005). This decision is complicated by the fact that this improved quality of life may come at a significant financial cost and reduced survival time.

This circumstance has been previously described by O'Connor et al., (2002) and is known as a choice dilemma or a conflicted decision (Sjoberg,

1983) characterized by uncertainty or difficulty in identifying the best alternative due to the risk or uncertainty of outcomes. The patient must make value judgments about potential gains versus potential loses (Keeney, 1982) and may anticipate regret over the positive aspects of rejected options (Sjoberg, 1983).

The literature review is discussed in Chapter 3 but to summarize we found only one study that explored informational needs of CF patients making the lung transplant decision and these authors concluded that, "the decision-making process could be facilitated and enhanced by practices that accommodate diverse informational needs in terms of content, quantity, timing and modality" (Moloney, Cicutto, Hutcheon, & Singer, 2007). There were no studies identified in the literature review that address how patients weigh the options of choosing referral for lung transplantation versus pursuing palliative care for their end-stage lung disease nor how this decision may change through time.

Decision Aids

Decision aids are used as adjuncts to counseling to prepare patients for decision making (O'Connor et al., 1999). They are different from educational materials which are generally broad in perspectives and help patients understand the treatment and management in general terms, but do not help the patients make a specific choice among options in an active, deliberate way (Coulter & Ellins, 2007, Winterbottom, Connor, Mooney, & Bekker, 2007).

Decision aids have been presented in a number of media formats including decision boards, audiotapes, audio-guided workbooks, interactive videodiscs, pamphlets, and personal computers (O'Connor et al.,1999). Some

aids are provided for patients to work through on their own and then used as a platform for discussion in consultation (Davison & Degner, 1997; Man-Son-Hing et al., 1999; O'Connor et al., 1998a) and others are delivered within consultations (Holmes-Rovner et al., 1999).

According to the Cochrane review, 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" (O'Connor et al., 1999, pg. 67) They may also include information on the disease, probabilities of outcomes, an explicit values-clarification exercise, information on others' opinions, and guidance or coaching in the steps of decision making and communication with others.

Decision aids are being developed in several centres in the United States, Europe, Australia and Canada (O'Connor et al., 2009). The trend is motivated by a) the rise of consumerism with an emphasis on informed choice rather than informed consent, b) evidence-based practice movement disseminating evidence to consumers as well as to practitioners, c) the use of consumer-based strategies to reduce practice variation, d) the realization that many decisions are "value laden" and depend on the values that patients place on benefits or risks and e) the increasing number of reviews and outcome studies that provide estimates of outcomes for use in decision aids (Elywn et al., 2006; O'Connor et al., 1998b; O'Connor et al., 2009).

A systematic review of 55 randomized controlled trials by O'Connor et al., 2009 concluded that decision aids improve patients' knowledge of the options,

create accurate risk perceptions of their benefits and harms, reduce decisional conflict, and increase active participation in the decision making process. However, there are no decision aids for patients with CF.

The Conceptual Framework

The Ottawa Decision Support Framework (ODSF) is a conceptual framework that was used to guide the development and evaluation of the lung transplantation decision aid. {Appendix D and Appendix E).

The ODSF is based on concepts from social psychology (Ajzen & Fishbein, 1980), cognitive psychology (Tversky & Kahneman, 1981) decisional conflict (Janis & Mann, 1977), decision analysis (Keeney, 1982) expectancy-value decision theories (Fischhoff, Slovic, & Lichtenstein, 1980), social support (Norbeck, 1988; Orem, 1995) and self-efficacy (Bandura, 1982).

The ODSF is an evidence based conceptual framework that is used to support patients in making difficult health or social decisions (O'Connor et al., 1998a). It recognizes the various participants involved in the choice including the individual, family, and health care practitioner. The ODSF describes a three step process which involves 1) assessing the client and practitioner needs or determinants of the decision, 2) providing decision support, and 3) evaluating the decision making process and outcome (O'Connor et al., 1998a).

According to the theories underlying the ODSF, determinants of decisions are factors that influence decisions and when sub-optimal lead to poor decision making (O'Connor, Jacobsen, & Stacey, 2002). Examples of sub-optimal determinants include:

1. Perceptions of the decision such as inadequate knowledge about the options, unrealistic expectations of benefits and risks, unclear values regarding personal importance of benefits and risks, and decisional conflict.

2. Perceptions of others such as inadequate support from others or pressure from others to choose one option.

3. Inadequate personal and external resources to make decisions (finances and limited skills in decision making).

4. Personal characteristics such as age, gender, ethnicity, education.

Decision support involves preparing the client and clinician for decision making and structuring follow-up counseling (O'Connor et al., 2002). The goal is to improve decision making by tailoring the support according to participants' needs. Examples of support strategies include:

1. Providing access to information about the clients' health situation, options and outcomes when a person's knowledge is inadequate.

2. Realigning expectations of the outcomes by describing them in detail including probabilities when a client's expectations are unrealistic.

3. Clarifying values by describing outcomes in detail using values clarification exercises so the client can better judge their values.

4. Providing information about others when a person has unclear or biased perceptions of others' opinions.

5. Helping the client access the support or resources needed (family, friends, health professionals) to make the decision.

6. Providing guidance or coaching in the steps of deliberating about a decision.

Evaluating the decision making process and outcome using the ODSF separates the evaluation of the decision making process from evaluation of outcomes because good decisions can result in bad outcomes but if decisions depend on the clients' values, the decision cannot be judged as right or wrong (O'Connor et al., 2002; O'Connor et al., 1998b). The ODSF defines a good decision as informed, consistent with personal values, acted upon and results in the client expressing satisfaction with decision making (O'Connor, 1995; O'Connor, 1997).

Rationale for Use of the Conceptual Framework

The Ottawa Decision Support Framework provided a practical and functional model to guide the assessment of the decisional support needs of patients with CF being referred for bilateral lung transplantation. The ODSF has guided the development and evaluation of several other decision aids including (Cranney et al., 2002 {osteoporosis}; Dales et al., 1999 {chronic obstructive pulmonary disease}; Fiset et al., 2000 {lung cancer}; O'Connor et al., 1998b {hormone replacement}, Stacey, O'Connor, DeGrasse, & Verma, 2003 {tamoxifien} and has also been used in previous health studies regarding decision making in populations facing life-threatening illness. (Stacey et al., 2003; Wilson et al., 2005).

The characteristics of the decision for CF patients re transplants is consistent with decisions best suited to the ODSF. These characteristics include: 1) there is no clear right choice; 2) it is stimulated by a new circumstance or

developmental transition; 3) it requires careful deliberation because of the value-laden outcomes; and 4) it requires more effort during the deliberation phase than the implementation phase (O'Connor, 1998a).

The ODSF is less useful for decisions whose key challenge is implementing and maintaining actions rather than making a choice and those that have no immediate stimulus for deliberation (O'Connor et al., 2002). For CF patients once the decision for transplant is made implementation is in control of the health care system not the patient.

Another strength of the ODSF is that it separates evaluation of the quality of the decision making from the outcomes of the decision. This is critical for this patient population as either choice (lung transplantation or usual care resulting in palliative care) can result in a bad outcome but the decision will depend on the patients' values and will not be judged as right or wrong (O'Connor et al., 2002).

Overall Summary

Patients with CF have progressive disease leading to increased morbidity and eventually death. Since 1988, double lung transplant has become available as a treatment option for CF patients with advanced disease. However, the decision to undergo transplant is complex given the need to weigh the potential benefits of transplant against the potential life threatening harms. Therefore, given the personal and complex decision making process for CF patients considering referral for lung transplantation, the ODSF was particularly suited to address this sensitive topic.

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Chapter Three

Literature Review, Needs Assessment, and Methods of Decision Aid Development

Chapter 3 is based on the following manuscript:

Vandemheen, K.L., Aaron, S.D., Poirier, C., Tullis, E., & O'Connor, A. Development of a decision aid for patients with cystic fibrosis considering lung transplantation. Accepted for publication to *Progress in Transplantation.*.

The chapter presents the comprehensive literature review, results of the needs assessment and the methods used to develop the decision aid. The references in the manuscript are formatted for the journal.

Contribution of Collaborators

There were five authors involved in the paper: Katherine Vandemheen (KV), Shawn Aaron (SA), Elizabeth Tullis (ET), Charles Poirier (CP), and Annette O'Connor (AO). KV wrote the first draft and SA and AO reviewed the draft and provided feedback. KV revised the paper and the revision was sent to SA, AO, ET and CP. All reviewed the paper and made editorial comments prior to its submission for publication.

All five authors contributed significantly to the development of the decision aid and funding was provided by The Ontario Thoracic Society and The Physicians' Services Incorporated Foundation.

Abstract

Context: Most adult patients with cystic fibrosis (CF) are eventually required to make a decision about referral for lung transplantation. Our objective was to identify the decisional needs of these patients and to develop a decision aid to address these needs.

Methods: We undertook a comprehensive review of the literature, a review of Canadian transplant statistics from 2002-2006, and a self-assessment survey of patients who had already made a decision about referral 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: We found a tremendous variation in transplant referral patterns amongst Canadian CF clinics. Canadian patients with CF who were not residing in transplant centers between 2002-2006 were significantly less likely to undergo lung transplants ($P < 0.0001$). Decisional needs identified by patients included wanting more information on; 1) relocation to the transplant centre; 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, we developed a lung transplantation decision aid. A panel of healthcare professionals and patients reviewed the decision aid and agreed the content was appropriate, easy to understand and unbiased.

Conclusion: The decisional needs of patients with CF who are considering lung transplantation are not being addressed in Canadian CF clinics, especially in

clinics outside of transplant centers. An evidence-based decision aid could serve as a useful tool to help address these needs.

Introduction:

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 the recipients (2). In Canada, five transplant centres perform bilateral lung transplantation on patients with cystic fibrosis. Since Canada is a geographically large country, many patients being referred for transplantation must move to the transplant city while they are waiting for their surgery. Furthermore, even after transplantation they must remain in the transplant city for 3-6 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 a very difficult decision for most patients and their families and they may require decision support.

In recent years there has been 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 there are no decision aids or evidence based tools 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 considering referral for lung transplantation were identified using a comprehensive review of the literature, a review of Canadian transplant statistics from 2002-2006 (6) and a self-assessment survey of patients from the Ottawa CF 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", "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 using the keywords "decision making", "decision tools", and "lung transplantation" and hand searches of journal articles were also conducted.

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

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 the perception 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 comprised of a nurse, two academic CF physicians/CF 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 CF decision aid followed the general format that is used for many of the other aids developed in Ottawa based on the Ottawa Decision Support Framework (7-9) and guided by the International Patient Decision Aid Standards as shown in Table 1 (10,11). 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 CF 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 CF health-care providers (2 physicians, 2 CF nurse coordinators) from across Canada and 4 CF patients who had already made a choice regarding referral for lung transplantation (one patient had declined referral for lung transplantation, one patient had just returned from their initial assessment of referral and two 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 (12,13). 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 two French Canadian CF clinicians. An

additional version, which presented alternative survival statistics, was created for patients chronically infected with *Burkholderia cepacia* since survival of these patients following lung transplantation differs from the general CF population (14).

Results:

The literature review identified one study that explored informational needs of CF patients making the lung transplant decision. Moloney and colleagues studied 22 CF patients who were transplant recipients or were awaiting lung transplant (15). The majority of transplant candidates and recipients stated that information on practical issues such as sources of financial assistance and relocation to the transplant centre were integral to making the transplant decision, yet these issues were inadequately addressed by the local CF clinic and by the transplant program. The authors of the study concluded that, "the decision-making process 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 (16,17).

The CF 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 there was very little information 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 there were 3782 Canadian patients followed in Canadian CF clinics. Of these 1547 patients (42%) received their care from a CF center located more than 2 hours from a lung transplant center. However, review of Canadian lung transplant statistics revealed that only 45 of 186 CF patients (24%), who received a lung transplant in Canada from 2002-2006 came from a CF center located more than 2 hours from a lung transplant center. Canadian patients with CF who were not residing in transplant centers between 2002-2006 were significantly less likely to have lung transplants ($P<0.0001$) compared to their counterparts who were residing within 2 hours of a lung transplant center.

Decision Aid Description:

The decision aid was developed as a paper-based booklet that is interactive, self-administered 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 in a young population who are computer savvy. The decision aid asks the patient to work through five steps when considering the options of "Not to be referred for lung transplantation" or "To be referred for lung transplantation". In step one the patient is asked to think about how CF affects them now. In step two they are asked to think about the options, benefits and risks of lung transplantation. The pros and cons of each option are presented using illustrative icons for the survival statistics and long term complications as seen in figure 1 (18). Step three of the decision aid asks them to choose the role they prefer in decision making. In step four they are asked to find out what else they need to prepare for decision making and in step five they are asked to plan the next steps once they have made their decision.

Questionnaire Results:

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

The four patients of 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 four patients felt the information provided was 'completely balanced', and one 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 stated that 'most things were clear'. All of the patients felt that the information material was 'very helpful' in helping them make a decision about the options and that they would 'definitely recommend' the material to other people facing the same decision.

The healthcare expert panel had similar responses with two people rating the information in the decision aid as having 'about the right information' and the other two rating it as 'a little less than needed' indicating that more information was needed on post-operative complications and long-term complications.

Three rated the length of the decision aid as 'just about right' and the other respondent thought it was 'a little too long'. Three healthcare providers felt the information provided was 'completely balanced' and one indicated that the information was a 'little slanted towards referral and lung transplantation'. All four healthcare providers indicated that the decision aid presented the 'options in a neutral and balanced manner'. All members of the healthcare panel felt that the information material was 'very helpful' in helping patients make a decision about

the options and that they would 'definitely recommend' the material to other patients facing the same decision.

Based on the results from the expert panel changes were made to the decision aid to include additional information about post-operative complications and a pictorial presentation was added to the survival statistics page which included risks of long term complications of diabetes, cancer and kidney failure.

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. (see figure 2) The terminology used in the decision aid was re-reviewed to ensure that it was consistent throughout the document.

Discussion:

This study aimed to identify the decisional needs of patients with cystic fibrosis being referred for bilateral lung transplantation. Once we identified specific needs we then developed a decision aid to meet these needs. Before development of our decision aid there were few decision support tools available to these individuals.

Our study showed that there is huge variation in referral patterns across Canada. Over 75% of CF patients who received lung transplants from 2002-2006 were referred from a CF clinic in close proximity to the transplant centre. Those patients who live distant 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, available to all over the internet, which presents information in an unbiased manner, would help inform patients with cystic fibrosis considering lung transplantation to better decide on their therapeutic options.

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

Our next step will be to formally assess if this decision aid will be a helpful adjunct in the decision making process. To do this we will conduct a randomized controlled trial of the decision aid vs. standard care. All patients in Canadian and Australian CF clinics whose lung function (measured as the forced expiratory volume in one second) is less than forty percent 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 CF physician and CF team immediately prior to randomization. Following 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 website which contains the interactive decision aid. Those randomized to 'usual care' will receive an information pamphlet from the Canadian CF Foundation which describes lung transplantation. All participants will complete standardized questionnaires before randomization and three 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.

Despite its limitations, the contribution of this study is important. Our study has assessed regional disparities in referral for lung transplantation, and has identified decisional needs of the CF community. The development of our decision aid is the first step forward towards providing a shared decision- making evidenced-based tool to a vulnerable population who are making what is arguably, the most important decision of their lives.

Acknowledgments:

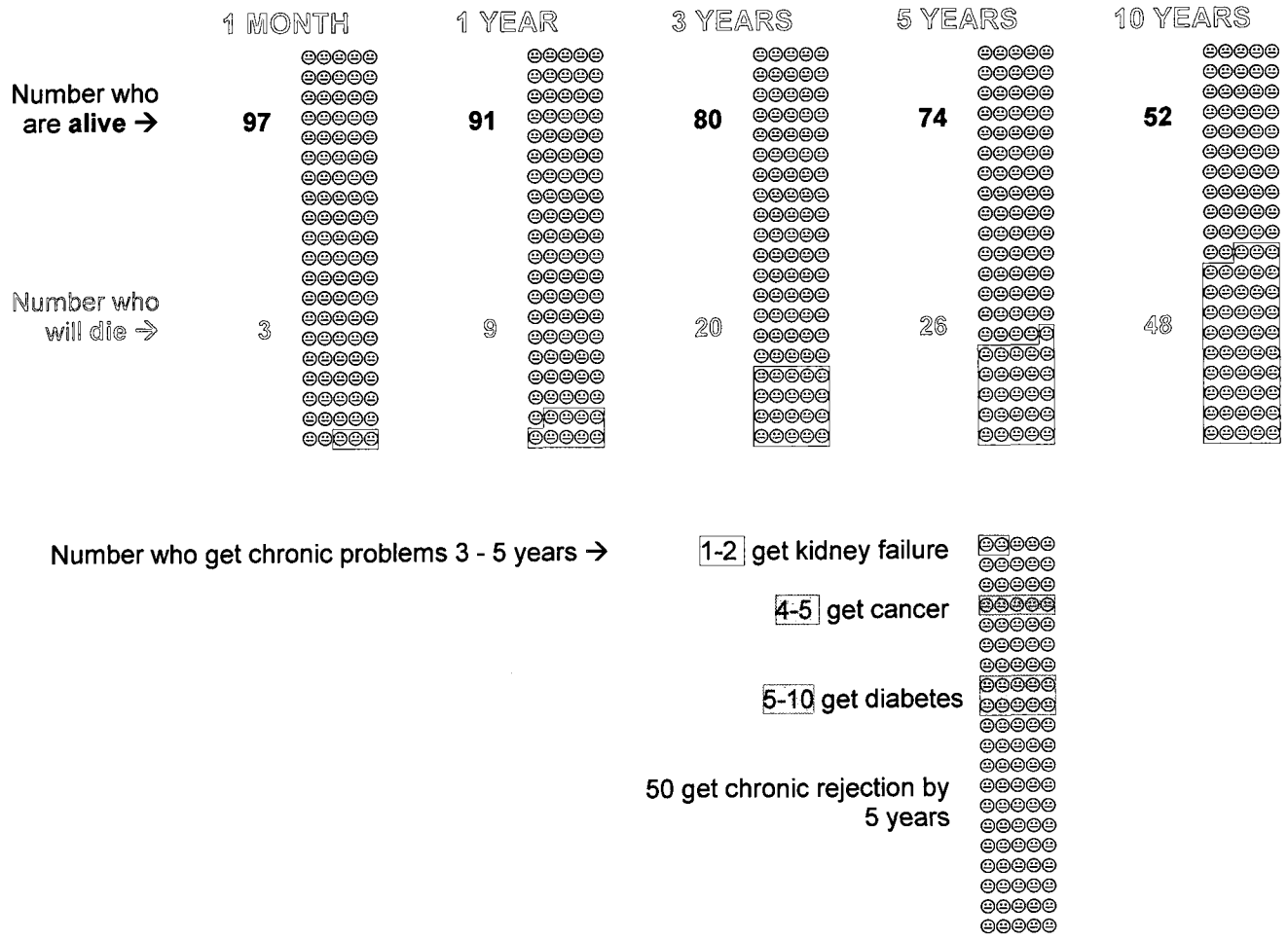
The authors would like to thank the cystic fibrosis patients and the CF specialists: Neil Brown MD, University of Alberta; Pearce Wilcox MD, University of British Columbia; Nancy Morrison MD, Dalhousie University; Patrice Kean BScN, London Health Science Centre; and Josette Salgado RN, University of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

Figure Legends:

Figure 1: Expected Results After Lung Transplantation as Presented in the Decision Aid

Figure 2: Benefits and Risks of Lung Transplantation as Presented in the Decision Aid

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 post transplantation.

Figure 2: Benefits and Risks of Lung Transplantation as Presented in the Decision Aid

1. Not to be referred for lung transplantation

- 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 laboured. 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**	○ 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 – 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**	○ 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 centre 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 ICU after surgery is 1-4 days** **Average time in hospital after surgery is 2-4 weeks**	○ Your new lungs will come from a person who has recently died and their 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 – 3 days. You will have tubes in your chest (chest tubes) and lines in your arms (intravenous) and wrist (arterial).
After hospital	**Average time is 3-6 months**	○ You will have to live in or very near your transplant center for several months after your transplant.

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.

Although your lungs will be healthier, you will still have CF. Lung transplant will not fix other CF health problems like diabetes, digestive problems, osteoporosis or male infertility.

Table 1. Methods for Developing Decision Aid according to the International Patient Decision Aid Standards (IPDAS)

IPDAS Standard [treatment]	Method
Is based on user's needs	**Patient Survey:** 3 patients who had made the decision at an Adult CF clinic using a semi-structured interview guide ***Record review:*** *Number of patient referrals and number of transplants linking to primary care CF centre*
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 CF, describes 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 centre while awaiting transplant, surgical complications and the possibility of organ rejection. 2. Probabilities presented as event rates over 10 year time period 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 CF adult patients, 4 CF Specialists reviewed the decision aid and completed the Acceptability questionnaire
Field testing shows the decision aid is acceptable to users	
There is evidence that the decision aid leads to informed, values-based decisions	In Progress, the decision aid is being evaluated in a randomized controlled trial.

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Chapter Four

Methods of Evaluation of the Decision Aid and Results of the Randomized Controlled Trial.

Chapter 4 is based on the following manuscript:

Vandemheen, K.L., O'Connor, A., Bell, S., Freitag, A., Bye, P., Jeanneret, A., Berthiaume, Y., Brown, N., Wilcox, P., Ryan, G., Brager, N., Rabin, H., Morrison, N., Gibson, P., Jackson, M., Paterson, N., Middleton, P., & Aaron, S.D. Randomized trial of a decision aid for cystic fibrosis patients considering lung transplantation. Accepted for publication to the American Journal of Respiratory and Critical Care Medicine submitted June 5, 2009.

The chapter presents the methods used in evaluating the decision aid in a randomized controlled trial in 149 patients in Canada and Australian CF clinics. The references in the manuscript are formatted for the journal.

Contribution of Collaborators

There were eighteen authors involved in the paper. Katherine Vandemheen (KV), wrote the first draft. Shawn Aaron (SA) and Annette O'Connor (AO) reviewed the draft and provided feedback. KV revised the paper and the revision was sent to the eighteen authors. All reviewed the paper and made editorial comments prior to its submission for publication.

Funding was provided by The Ontario Thoracic Society, The Physicians' Services Incorporated Foundation and The Australian Cystic Fibrosis Research Trust.

Abstract

Rationale: We developed an evidence based decision aid for cystic fibrosis patients with advanced lung disease considering referral for lung transplantation.

Objective: To prospectively evaluate whether use of the decision aid increased knowledge about the options, improved realistic expectations, and decreased decisional conflict in adult patients.

Methods: We performed a single-blind randomized controlled trial involving 149 adult cystic fibrosis patients with an $FEV_1 \leq 40\%$ predicted from 14 Canadian and Australian centers. All participants received an education and counseling session from their cystic fibrosis team and were then randomized to receive the decision aid or usual care.

Measurements and Main Results: The primary end points measured were participants' knowledge, realistic expectations, and decisional conflict evaluated three weeks after randomization. Patients randomized to the decision aid had greater knowledge about their options ($P < 0.0001$) and had more realistic expectations about the benefits and risks of lung transplantation ($P < 0.0001$) compared to those randomized to usual care. The total decisional conflict score was significantly lower in the decision aid group 3 weeks post randomization compared to the usual care group (11.6 vs 20.4; $P = 0.0007$). Decisions were durable; 88% of patients in the decision aid group and 75% in the usual care group maintained the same choice 12 months after randomization ($P = 0.06$).

Conclusions: Use of a decision aid for cystic fibrosis patients considering referral for lung transplantation, in addition to usual education and counseling,

improves patient knowledge, realistic expectations, decisional conflict, and patient satisfaction.

Scientific Knowledge on this Subject

This is the first patient decision aid to be used in the area of cystic fibrosis and the first in the area of lung transplantation.

What the study adds to the Field

This randomized, controlled trial demonstrated that use of a decision aid improved patient knowledge, realistic expectations, decisional conflict, and patient satisfaction in adults with CF considering lung transplantation.

This article has an online data supplement, which is accessible from this issue's table of content online at www.atsjournals.org and in Appendix O

This study is registered with Clinical Trials.gov sponsored by the National Institutes of Health number NCT00345449.

Introduction

Cystic Fibrosis (CF) is one of the most common inherited fatal diseases. Over the past forty years, the median age of survival has improved from fourteen years in 1969 to age thirty-seven in 2007 in Canada and the US [1]. With this improved survival, CF has been transformed from a disease with very high infant mortality to a disease that must now be considered progressive and chronic. However eventually most patients with CF develop end-stage bronchiectasis and obstructive lung disease that cannot be improved with conventional medical therapy.

When conventional medical regimens fail and disease progresses in patients with severe lung disease, lung transplantation can be the only option left. For the appropriate candidate, lung transplantation can improve pulmonary function, exercise tolerance, quality of life and survival [2], but lung transplantation nonetheless remains an imperfect technique. The risk of infection and rejection are ever-present and survival beyond 5 years is realized by only half of the recipients [3]. Of these survivors 50-60% suffer from bronchiolitis obliterans syndrome leaving them with potentially significant morbidity [4].
Some recent studies have suggested potential decreased survival with lung transplantation in children with CF but no randomized controlled trial has ever been done to assess lung transplantation versus usual care [5].

Previous studies of CF patients and providers have identified that patients with CF experience considerable anxiety and uncertainty related to making decisions concerning whether to be referred for lung transplantation [6;7]. In

response to this we developed an evidenced- based decision aid for cystic fibrosis patients with severe lung disease who were considering referral for lung transplantation. The methods of the development of the decision aid have been previously described and submitted for publication [6].

The purpose of this randomized controlled clinical trial was to determine whether use of the decision aid improved the quality of decision making of cystic fibrosis patients when considering referral for lung transplantation in comparison to usual care. The primary objectives were to assess if the decision aid increased knowledge about patient options, improved realistic expectations, and decreased decisional conflict. The secondary objectives were to assess the patients' satisfaction with the decision aid, preparation for decision making, and the patients' stated choice among the options. Durability of the decision was also measured twelve months after randomization.

Methods

Patients

We enrolled patients with CF from 9 Canadian and 5 Australian outpatient centers. Eligible patients had to be 18 years of age or greater, have a forced expiratory volume in one second (FEV_1) less than or equal to 40% predicted, and be able to read English or French. Patients who had previously received a lung transplant, and those on lung transplant waiting lists, were excluded. The research ethics boards of all the participating centers approved the study, and all trial participants provided written informed consent.

Study Intervention

This was a single blind randomized controlled trial with a prospective one year follow-up. All study patients received an education session on the process of referral and the risks and benefits of lung transplantation from their CF physician and/or CF team. The local CF clinic staff and physicians were also encouraged to distribute any other material normally provided to patients considering lung transplant to all participating patients. All patients received a copy of a pamphlet entitled, "Cystic fibrosis and Lung Transplantation" [8] and were provided addresses to access two generic websites on lung transplantation. Prior to randomization, all study patients completed standardized questionnaires about knowledge of the options, expectations, decisional conflict and preference for referral and lung transplantation.

Randomization was through central allocation of a randomization schedule prepared through a computer-generated random listing of the two treatment allocations blocked in variable blocks of two or four and stratified by site and by infection status with *Burkholderia cepacia*. Neither research staff nor medical staff was aware of the treatment assignment before or after randomization.

Patients were randomized after receiving their lung transplant educational session and after filling out the baseline questionnaires. Those in the decision aid arm received a sealed package containing a paper version of the decision aid and a website address with a user name and password where they could access the decision aid on-line. If the patient was randomized to the usual care arm the package contained blank pages and a letter explaining why blank pages were

included. Patients were instructed not to open the packages until arriving home and they were told not to inform the research staff of their randomization status.

Three weeks after randomization the research staff who were blinded to treatment allocation telephoned each patient and had them complete a follow-up questionnaire. The follow-up questionnaire was similar to the one presented at baseline and assessed knowledge about lung transplantation, the patient's expectations, decisional conflict, and whether they had made a choice to be referred, or to forego, lung transplantation.

The development of the CF Decision aid is described in the on-line data supplement (Appendix O). A web-based version of the decision aid is available online at http://decisionaid.ohri.ca/decaids.html

Outcome Measures

The primary end points were participants' knowledge, realistic expectations, and decisional conflict scores. All three primary outcomes were compared between the two treatment groups three weeks after randomization. Knowledge was measured using a 4-item multiple choice questionnaire designed to determine the patients' understanding of the benefits and risks of referral and lung transplantation, or of foregoing referral. The response categories were coded as correct or incorrect. The items were summed and converted to a mean score (range of 0 to 4). Realistic expectations evaluated the patients' perceptions of the level of risk for surgery and the probabilities of survival with and without surgery. Responses were coded as correct or incorrect and mean scores were calculated (range 0 to 2). Both the knowledge and realistic expectations

questionnaires followed a structured format as per the International Patient Decision Aids Standards Guidelines [9;10]. Further details concerning these questionnaires can be found in the online data supplement.

Decisional conflict is defined as: "uncertainty about the course of action to take when choice among options involves risk, loss, regret or challenge to personal life values"[11]. Decisional Conflict was evaluated using the Decisional Conflict Scale, a validated 16 item scale with 5 subscales. All questions use a five item response scale. Scores vary from 0 (low decisional conflict) to 100 (high decisional conflict). The instrument has a minimal clinically important difference previously established; patients scoring 37.5 or more are likely to delay decisions, whereas those with a score of 25 or less tend to make decisions[11].

Secondary outcomes included patient preparation for decision making, choice, and durability of the decision. Patients were asked to indicate whether a decision regarding the choice of referral for lung transplantation had been made at baseline and three weeks after randomization. Durability of the decision was assessed twelve months later by telephone. Between-group differences in changes in knowledge, realistic expectations, and decisional conflict scores from week 3 to baseline were also assessed as secondary outcomes.

Other secondary outcomes assessed included value congruence with the choice made and acceptability of the decision aid and the materials provided [12]. Additional detail on the assessment and evaluation of these outcome measurements is provided in an online data supplement.

Statistical analysis

We calculated the sample size required to detect the minimal clinically important differences between the decision aid group and the usual care group for the three outcomes; knowledge, decisional conflict and expectations. Previous studies have shown that an effect size of 0.5 for these 3 outcomes is considered minimally important since this difference is able to discriminate between patients who do and do not make decisions. The significance level of 0.05 was adjusted to 0.0167 using a Bonferroni correction to accommodate the testing of the three major outcomes. Using expected standard deviations for decisional conflict, expectations and knowledge of 0.6, 22.5, and 18 respectively [13-15], an expected effect size of 0.5, and a power of 80%, we determined that 100 randomized patients were required for each group.

Descriptive statistics with 95% confidence intervals were used to compare baseline characteristics for the two randomized groups. Chi-square tests were used to compare the proportions of patients who answered knowledge and realistic expectations questions correctly. Secondary subgroup analyses were performed for all outcomes stratifying results by gender. The decisional conflict scores and preparation for decision making were compared between groups using independent t-tests. In addition, linear regression adjusting for age, gender, and FEV_1 % predicted was used to compare the between group change (week 3 score – baseline score) and the 3-week preparation for decision making scores. Values congruence was evaluated using logistic regression analysis for the outcome 'choosing lung transplantation' using the answers to the 4 values

questions as predictors. Separate regressions were run to assess values congruence before and after randomization to the intervention. All analyses were intention to treat and were conducted using SAS software, version 9.0 (SAS Institute, Inc., Cary, North Carolina).

Results

Study Population

Patients were randomized from November 2006 to June 2008. A total of 155 eligible patients were screened for the study however six patients declined to participate. One hundred and forty-nine patients (92 from Canada and 57 from Australia) met the eligibility criteria and agreed to participate in the study and were randomized to either receive (n=70) or not receive (n=79) the decision aid (Figure 1). Recruitment was stopped after 149 patients were randomized once all of the potentially eligible patients in the 14 centers had been approached for entry into the trial. One patient in the control group dropped out of the study and did not complete the three week telephone follow-up questionnaire.

The baseline demographics and clinical characteristics of the two groups are provided in Table 1. The FEV_1% predicted in the decision aid group was 32% (SD 5.8) vs 31% (SD 6.8) in the usual care group. Randomization resulted in a gender imbalance with more males being randomized to the decision aid group compared to the usual care group (64% of patients in the decision aid group were male vs 46% in the usual care group). The two groups had similar medication profiles and exacerbation rates. Level of education was self reported by the patient and was similar in both groups. Patients in both randomized

groups spent a similar amount of time reviewing the presented materials on lung transplantation (Table 7). Only 7 of the 70 patients randomized to the decision aid group accessed the on-line version, most worked from the paper copy.

Primary outcome

Knowledge

Patient knowledge scores at baseline in the two groups were similar (P= 0.79). However at 3-week follow-up those who were randomized to the decision aid had a significantly higher proportion of correct scores on the knowledge questionnaires compared to patients who received usual care (P < 0.0001) (Figure 2). In addition, the three week change in mean knowledge scores was significantly higher in the decision aid group (1.24 (SD1.38) compared to the control group (0.30 (SD 1.17); P < 0.0001 (Table 2). Adjustment of the results for gender had no effect on the results; both males and females randomized to the decision aid had significantly higher knowledge scores compared to those randomized to usual care.

Expectations

Patient expectation scores in the two groups were similar at baseline before randomization (Table 3). Three weeks after randomization, those patients in the decision aid group were significantly more likely to provide correct quantitative estimates of their risk of death when choosing or foregoing lung transplantation in comparison to the usual care group. In addition, the three week change in mean expectation scores was significantly higher in the decision aid group (0.71 (SD 0.98) compared to the control group (0.05 (SD 0.81); P < 0.0001.

Decisional Conflict

Decisional conflict scores were not significantly different between the two groups at baseline (total baseline decisional conflict score was 27.9 (SD 23.7) in the decision aid group versus 33.9 (SD 23.7) in the usual care group; P = 0.13 (Table 4). At week three the decision aid group had significantly lower total decisional conflict scores compared to the usual care group (11.6 (SD 13.6) versus 20.4 (SD 16.9); P = 0.0007 (Figure 3). Scores for all 5 decisional conflict subscales at week three were also significantly lower in those patients randomized to the decision aid (Table 4). Post-hoc linear regression adjusting for age, gender, FEV_1 % predicted, and the patient's baseline total decisional conflict score similarly showed that patients who were randomized to the decision aid had significantly lower decisional conflict scores at 3 weeks compared to the control group; the adjusted mean difference in total score between treatment groups was 6.0 (95% CI; 2.2-9.8; P = 0.003). Decisional conflict scores after receiving the information materials did decrease for subjects in both groups such that the absolute unadjusted change in decisional conflict over the three week period was not significantly greater for those randomized to the decision aid (16.4 (SD 17.9) vs those randomized to usual care (13.1 (SD 16.7); P = 0.25 (Table 4).

Secondary outcomes

Patient's Stated Choice

Prior to randomization, a similar proportion of patients in both groups expressed preference for referral for lung transplantation, preference for no referral, or were unsure. Randomization to either the decision aid or usual care

group did not significantly influence the eventual choice made by the patient (P=0.77) (Table 5).

Durability of the Patient Decisions

Seven of 149 patients (5%) died, and 13 patients (9%) had a double lung transplant within 12 months of randomization (Figure 1). Four patients were lost to follow-up at 12 months or prematurely withdrew from the study (2 in each randomized group). Of the remaining 125 randomized patients, 53/60 patients (88%) in the decision aid group, and 49/65 patients (75%) in the usual care group chose the same option (transplant, no transplant, or unsure) as they had chosen 3 weeks after randomization into the study. There was no significant difference in durability of choice between the two groups; P=0.06.

Preparation for Decision Making

A questionnaire evaluating preparation for decision making was administered at the three week follow-up. Patients in the decision aid group felt that the information prepared them more thoroughly for making a decision than patients in the usual care group. The overall mean preparation for decision making score was 65.1 (SD 24.9) in the decision aid group compared to 53.8 (SD 27.1) in the usual care group (mean difference = 11.3; P=0.009) (Table 6).

Values Congruence

Patient choices were consistent with their values across both randomized groups. Those patients who chose referral and lung transplantation rated living longer as very important (odds ratio = 1.80; 95% CI; 1.02 - 3.17; P=0.04) and placed less importance on avoiding the hassle, stress and worry of lung

transplantation (odds ratio = 0.77; 95% CI; 0.6-1.00; P=0.047) compared to those who declined lung transplantation. .

Acceptability

Patients' ratings for acceptability of the information materials are shown in Table 7. Seventy four percent of the patients in the decision aid group reported that they received about the right amount of information in comparison to 39% in the usual care group (P<0.001). The decision aid group was more likely to report that the information materials were very helpful in helping them arrive at a decision (47% vs 35%; P=0.01). More patients randomized to the decision aid group stated they would definitely or probably recommend the materials compared to those randomized to usual care (97% vs 87%; P=0.03).

Posthoc Subgroup Analysis: Effects of Education

To determine if there was any effect of the patient's educational attainments on the effectiveness of the intervention we divided those patients who were randomized to the decision aid into those who graduated high school or less (N=34) and those who completed at least some college or higher (N =36). Knowledge scores at 3 weeks were higher in the group with higher education (total knowledge score = 3.36 (SD 0.90) vs 2.53 (SD 1.11), P = 0.0009). However total decisional conflict score at 3 weeks was not different in the two educational groups (11.4 (SD 13.8) vs. 11.7 (SD 13.6), P = 0.94). There was also no significant change in the preparation for decision making score between the 2 educational groups (60.7 (SD 26.5) vs 69.4 (SD 22.9), P=0.14).

Discussion

Our study found that patients with advanced CF lung disease who were randomized to a lung transplant decision aid had a considerable increase in knowledge and had more realistic expectations about their level of risk for surgery and the probabilities of survival with and without surgery compared to those who received usual care and counselling. A previous study by Moloney et al reported that recipients of lung transplants who experienced a protracted recovery period following surgery expressed regret about undergoing transplantation, stating that they would have "benefited from being more prepared for their post-transplant experiences" [7]. Clearly, creating realistic expectations before the transplant procedure may have value beyond its effect on the decision itself. Use of the decision aid in our study was shown to improve realistic expectations of the potential complications post lung transplant and might be expected to better prepare patients for the post-transplant experience.

Patients in our study had somewhat lower baseline decisional conflict scores compared to baseline decisional conflict scores reported by patients facing other healthcare decisions in other studies [16;17]. Previous studies have shown that patients with a total decisional conflict score above 38 tend to delay decisions, whereas those with a score of 25 or less tend to make decisions [14;18]. Therefore, prior to being randomized into our study clearly some patients had already been ready to make a decision. This likely occurred because we chose patients with advanced CF lung disease for our study. Most CF patients have been ill since childhood with a chronic disease, they receive their health care in

specialized CF clinics, and they are thus relatively well informed about their disease and the eventual need to make a decision about lung transplantation. Even though patients in our study had lower than expected baseline decisional conflict scores, our study was still able to demonstrate significant differences in overall decisional conflict, and lower 3-week decisional conflict scores in the group that received the decision aid. In addition, patients who received the decision aid reported being more informed, more certain about their choice, had clearer values and were more likely to report satisfaction with their decision.

As patients have become "effective consumers" they increasingly want to make their own decisions [19], and it is crucial that information be presented to them in an unbiased manner. More than half the patients who received the decision aid rated the material as completely balanced. A similar proportion of patients in both the decision aid and the usual care arms chose to be referred for transplant or chose to decline transplant. This indicates that use of the decision aid helped patients to make a choice, but did not sway them towards one direction or the other.

This study has some limitations that should be considered. All participants received a counseling session and standardized written material about lung transplantation prior to randomization. Questionnaires on acceptability of the provided material indicated that the written material provided to the control group was well-received and thus may have reduced the impact of the decision aid.

A second potential limitation is the sample size. We had planned to randomize 200 patients into the trial but we stopped recruitment at 149 patients

once all of the potentially eligible patients in the 14 centers had been approached for entry into the trial. Although the primary outcomes which were measured at 3-weeks were all statistically positive in favor of the decision aid group, the change in the overall decisional conflict score (a secondary outcome), favored the decision aid group, but was not statistically different compared to the change in the overall decisional conflict score in the control group. It is possible that if we had recruited more patients into the trial then this difference would have become statistically significant.

Finally, we did not stratify randomization by gender, and by chance more male patients were randomized to the decision aid group then to usual care group (64% vs 46%) creating a gender imbalance. However, adjustment of the results for gender had no effect on knowledge scores, realistic expectation scores or decisional conflict scores.

The strengths of this study are its randomized design, and its multicentre international focus. Control patients, who received individual counseling sessions and written pamphlets on lung transplantation provided by the Canadian CF Foundation[8], were arguably provided with a better than standard-of-care approach. The fact that the decision aid group was able to demonstrate better outcomes despite the smaller sample size and despite the relatively excellent materials provided to the control group argues for the effectiveness of the decision aid as a patient care tool.

To our knowledge this is the first decision aid to be used in the area of CF and the first in the area of lung transplantation. We believe that this decision aid

can be incorporated easily into the general practice of CF. Most Canadian, American, Australian and European CF clinics have a dedicated team of healthcare providers who care for these patients, and the nurses within these teams have expertise in patient teaching. These specialized care nurses are well positioned to take an active role in supporting these patients with their decision making. The same design that was used in this study can be used in everyday practice with the patient receiving an education session on the process of referral and the risks and benefits of lung transplantation from their CF physician and then receiving a copy of the decision aid or the web address where they can view it on-line. Follow-up discussion after review of the decision aid can occur at the next patient visit.

In summary, this study has demonstrated a positive impact of a decision aid for cystic fibrosis patients considering referral for lung transplantation. It is hoped that this free, convenient, and easily accessible tool will be incorporated into CF patient care.

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Data managers: My-Linh Tran and Jennie Cote.

Statistician: Steve Doucette

Figure and Table Legends:

Figure 1: **Trial Profile**

Figure 2: **Knowledge Scores.**

The proportion of patients who answered all four knowledge questions correctly is shown at baseline and 3-weeks post randomization

Black bar = Decision Aid; Grey bar = Usual Care

Figure 3: **Overall Decisional Conflict Scale Score at Baseline and 3-weeks post randomization**

Black bar = Decision Aid; Grey bar = Usual Care

Table 1: **Base-Line Characteristics of the 149 Randomized Patients According to Treatment Assignment.**

Table 2: **Patient Knowledge Mean Scores at Baseline and 3-week Follow-up.**

Table 3: **Realistic Expectation Scores at Baseline and 3-week Follow-up.**

Table 4: **Decisional Conflict Scale Scores at Baseline and 3-week Follow-up**

Higher scores indicate greater decisional conflict. Scores vary from 0 (low decisional conflict) to 100 (high decisional conflict)

Table 5: **Patients' Stated Choice Before and After Randomization**

Table 6: **Preparation for Decision Making**

Table 7: **Acceptability of the Decision Aid**

Figure 1: Trial Profile

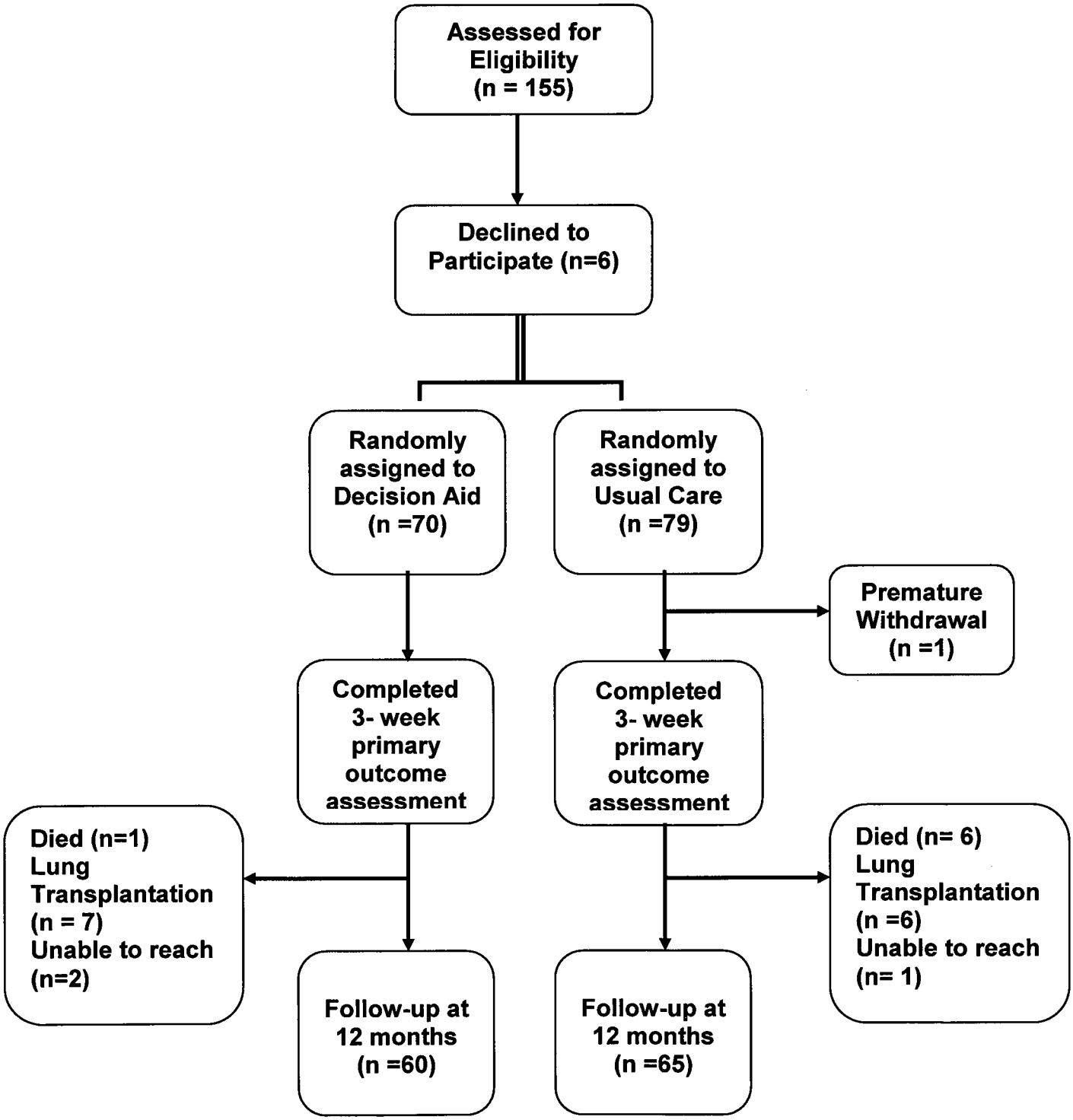

Figure 2:

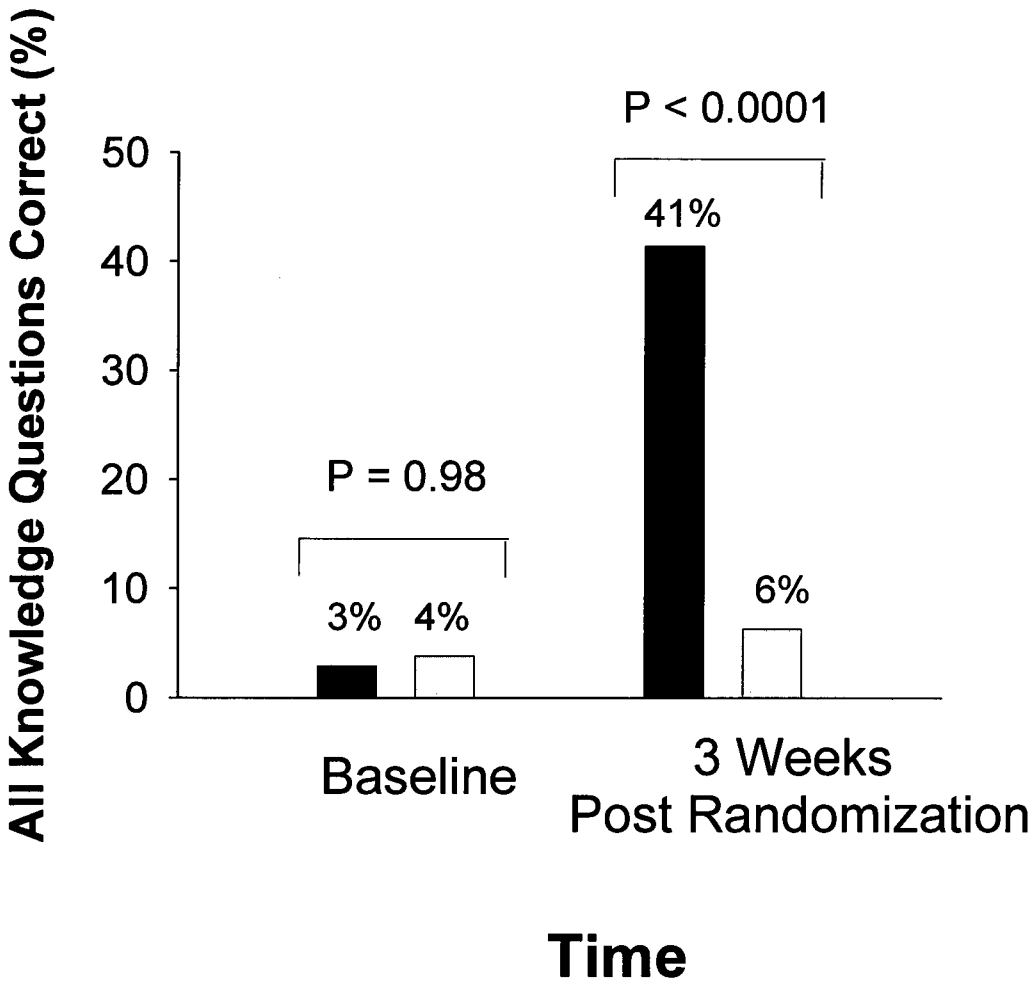
All Knowledge Questions Correct (%)
50
40
30
20
10
0
P < 0.0001
41%
P = 0.98
3%
4%
6%
Baseline
3 Weeks
Post Randomization
Time

Figure 3:

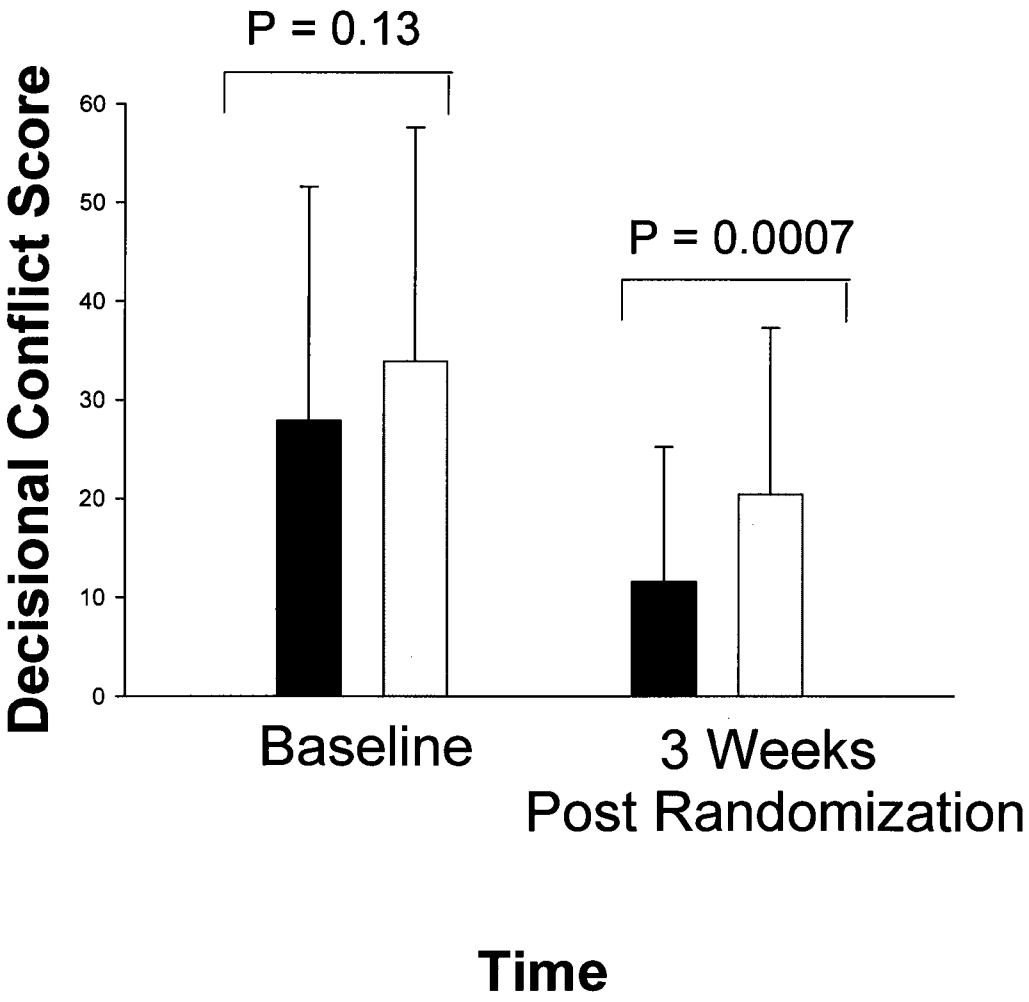
P = 0.13
P = 0.0007
Decisional Conflict Score
60
50
40
30
20
10
0
Baseline
3 Weeks
Post Randomization
Time

Table 1: Baseline Characteristics of the 149 Randomized Patients according to treatment assignment:

Characteristic	Decision Aid (N =70)	Usual Care (N = 79)
Age	30.1 ± 9	30.7 ± 9
Male (%)	64	46
Body Mass Index (kg/m^2)	20.4 ± 2.5	20.3 ± 2.9
FEV1 (L)	1.20 ± 0.25	1.10 ± 0.26
FEV1 % predicted	31.5 ± 5.8	30.6 ± 6.8
FVC (L)	2.5 ± 0.65	2.2 ± 0.69
FVC % predicted	55.7 ± 11.9	52.8 ± 14.5
Medications (%)		
Tobramycin or TOBI	47	52
Pulmozyme	63	47
Colistin	31	27
Azithromycin (chronic)	70	73
Medical Conditions (%)		
CF-related Diabetes	23	35
Pancreatic Insufficiency	90	94
CF-related Liver Disease	17	9
Organisms in Sputum (%)		
Burkholderia cepacia	9	9
Pseudomonas aeruginosa	84	84
Stenotrophomonas maltophilia	19	15
Staphylococcus aureus	49	41
Number of Exacerbations past year (%)		
None	6	8
1	17	9
2-3	30	35
4-5	27	23
>5	20	25
Number of Exacerbations past year requiring admission (%)		
None	31	27
1	24	24
2-3	30	35
4-5	9	10
>5	6	4

Number of Days in hospital (%)		
None	29	25
1-30	37	42
> 30	34	33
Highest Level of Education Attained (%)		
Graduated university or college	42	42
Attended university or college but did not graduate	9	14
Graduated high school	20	27
Attended high school but did not graduate	24	14
8^{th} grade or less	4	4

Table 2: Patient Knowledge Scores at Baseline and 3-week Follow-up

Knowledge Questions (maximum score =4)	Decision Aid (N =70) Baseline	Usual Care (N = 79) Baseline	Between group difference (95% CI) P value)	Decision Aid (N =70) 3-weeks	Usual Care (N = 79) 3-weeks	Between group difference (95% CI) P value)
Mean Score (SD)	1.71 (3.95)	1.67 (4.40)	0.04 (-0.27, 0.36) P = 0.79	2.96 (4.50)	2.00 (3.26)	0.96 (0.66, 1.31) P<0.0001
Change in score (week 3 – baseline)				1.24 (1.38)	0.30 (1.17)	0.94 (0.53, 1.35) P<0.0001

Table 3: Realistic Expectation Scores at Baseline and 3-week Follow-up

Realistic Expectations (maximum score =2)	Decision Aid (N =70) Baseline	Usual Care (N = 79) Baseline	Between group difference (95% CI) P value)	Decision Aid (N =70) 3-weeks	Usual Care (N = 79) 3-weeks	Between group difference (95% CI) P value)
Mean Score (SD)	0.60 (0.73)	0.53 (0.70)	0.07 (-0.16, 0.30) P = 0.79	1.31 (0.75)	0.58 (0.63)	0.73 (0.51,0.96) P<0.0001
Change in score (week 3 – baseline)				0.71 (0.98)	0.05 (0.81)	0.66 (0.37,0.95) P<0.0001

Table 4: Decisional Conflict Scale Scores at Baseline and 3-week Follow-up

Characteristic	Decision Aid (N =70)	Usual Care (N = 79)	Between group difference (95% CI) P value)	Decision Aid (N =70)	Usual Care (N = 79)	Between group difference (95% CI) P value)
	Baseline	Baseline	Baseline	3-weeks	3-weeks	3-weeks
Total Score (maximum score =100)	27.9 ± 23.7	33.9 ± 23.7	6.0 (-1.7, 13.7) p=0.13	11.6 ± 13.6	20.4 ± 16.9	8.8 (3.8, 13.9) p=0.0007
Subscale (maximum score =100 for each subscale)						
Certainty	39.6 ± 33.5	45.4 ± 33.2	5.7 (-5.1, 16.5) p=0.30	26.4 ± 25.9	36.4 ± 27.8	10.0 (1.3, 18.8) p = 0.03
Informed	30.5 ± 35.0	37.3 ± 32.6	6.9 (-4.1, 17.8) p=0.22	4.5 ± 9.6	17.2 ± 20.6	12.7 (7.4, 18.0) p<0.0001
Values	33.5 ± 34.6	38.5 ± 32.9	5.1 (-5.9, 16.0) p=0.36	9.9 ± 17.7	16.8 ± 21.0	6.9 (0.6, 13.2) p=0.03
Support	15.1 ± 17.1	22.4 ± 17.4	7.2 (1.6, 12.9) p=0.01	6.9 ± 12.3	14.5 ± 17.7	7.6 (2.6, 12.6) p=0.003
Satisfaction with Decision	22.7 ± 21.4	28.0 ± 26.0	5.3 (-2.4, 13.1) p=0.18	10.4 ± 16.4	17.9 ± 20.4	7.5 (1.5, 13.6) p=0.02
Change in Total Score (week 3 – baseline)				16.4 ± 17.9	13.1 ± 16.7	-3.3 (-8.9, 2.3) p= 0.25

Table 5: Patients' Stated Choice Before and After Randomization

	Decision Aid Visit #1	Usual Care Visit #1	Decision Aid 3-weeks	Usual Care 3-weeks
Referral for Transplant	35 (50%)	41 (53%)	47 (67%)	55 (70%)
Declined Referral for Transplant	13 (19%)	8 (10%)	10 (14%)	7 (9%)
Unsure	22 (31%)	29 (37%)	13 (19%)	16 (21%)

Table 6: Preparation for Decision Making

Characteristic	Decision Aid (N = 70) Maximum score = 100	Usual Care (N = 79) Maximum score = 100	Between group difference (95% CI) P value)
	Visit 3	Visit 3	
Did the information help you recognize that a decision needs to be made?	64.6 ± 30.8	53.8 ± 32.3	10.8 (0.5, 21.1) p=0.04
Did the information prepare you to make a better decision?	62.9 ± 32.3	52.2 ± 32.3	10.6 (0.1, 21.1) p=0.048
Did the information help you think about the benefits and risks of each option?	72.5 ± 29.5	58.3 ± 32.9	14.2 (4.0, 24.4) p=0.007
Did the information help you think about which benefits and risks are most important?	70.0 ± 29.4	50.6 ± 32.2	19.4 (9.3, 29.4) p=0.0002
Did the information help you know that the decision depends on what matters most to you?	70.7 ± 29.8	53.8 ± 33.7	16.9 (6.5, 27.3) p=0.002
Did the information help you organize your own thoughts about the decision?	57.9 ± 31.7	52.6 ± 30.6	5.3 (-4.8, 15.4) p=0.30
Did the information help you think about how involved you want to be in this decision?	63.9 ± 34.0	57.7 ± 34.7	6.2 (-4.9, 17.4) p=0.27
Did the information help you identify the questions you want to ask the doctor?	64.3 ± 28.7	54.5 ± 30.9	9.8 (0.07, 19.5) p=0.048
Did the information prepare you to talk to your doctor about what matters most to you?	65.7 ± 27.6	52.9 ± 31.5	12.8 (3.2, 22.5) p=0.01
Did the information prepare you for a follow-up visit with your doctor?	58.9 ± 34.1	51.6 ± 34.5	7.3 (-3.8, 18.5) p=0.20
Total Score	65.1 ± 24.9	53.8 ± 27.1	11.3 (2.8, 19.8) p=0.009

Table 7: Acceptability of the Decision Aid

Characteristic	Decision Aid (N =70) %	Usual Care (N = 79) %	*P*-Value
Did you read all the information material that you were provided?			
No, I did not read any of the information	**0**	**1**	**0.34**
Yes, I read it all	**90**	**94**	
I read some of the information	**10**	**5**	
How long did you spend reading and thinking about the information?			
Less than 1 hour	**36**	**36**	**0.53**
Between 1 and 6 hours	**52**	**46**	
More than 6 hours	**12**	**18**	
How would you rate the amount of information in the materials you were provided?			
Much less than I needed	**1**	**24**	**<0.0001**
A little less than I needed	**17**	**27**	
About the right amount of information	**74**	**39**	
A little more information than I needed	**6**	**7**	
A lot more information than I needed	**1**	**3**	
How balanced was the information in the material you received?			
Clearly slanted towards referral and lung transplantation	**10**	**21**	**0.24**
A little slanted towards referral and lung transplantation	**39**	**38**	
Completely balanced	**51**	**39**	
A little slanted towards no referral for lung transplantation	**0**	**1**	
Clearly slanted towards no referral for lung transplantation	**0**	**1**	
How clear was the information in the material?			
Everything was clear	**54**	**37**	**0.22**
Most things were clear	**44**	**56**	
Some things were clear	**1**	**3**	
Many things were unclear	**1**	**4**	
How helpful was the information material in helping you made a decision about options?			
Very helpful	**47**	**35**	**0.01**
Somewhat helpful	**38**	**33**	
A little helpful	**12**	**19**	
Not helpful	**3**	**13**	

Would you recommend these materials to other people facing same decision?			
I would definitely recommend it	**57**	**63**	**0.03**
I would probably recommend it	**40**	**24**	
I would probably not recommend it	**3**	**7**	
I would definitely not recommend it	**0**	**6**	

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Chapter Five
Discussion with Implications for Nursing

This chapter integrates the findings of the decisional needs of cystic fibrosis patients considering referral for lung transplantation and the results of using the decision aid in a prospective trial. Contributions of the study to the discipline of nursing and implications for practice, education, policy and research completes the chapter.

Discussion

In this thesis, the decisional needs of patients with CF with advanced lung disease who were considering referral for bilateral lung transplantation were identified and a decision aid to meet these needs was developed and evaluated in 149 patients. The main results of the needs assessment indicated that Canadian transplant statistics from 2002-2006 showed a wide variation in transplant referral patterns amongst the Canadian CF clinics; patients with CF who were not residing in transplant centers were significantly less likely to undergo lung transplants ($P < 0.0001$). (Chapter 3) The survey with Ottawa patients who had already made a decision about referral revealed decisional needs which included wanting more information on; 1) relocation to the transplant centre; 2) the benefits and risks of surgery; and 3) how to cope with anxiety and depression when making the decision. (Chapter 3)

In response to patients' information needs, we developed the lung transplantation decision aid based on the Ottawa Decision Support Framework

(O'Connor et al., 1998) and guided by International Patient Decision Aid Standards (Elwyn et al., 2006). Elements included: facts about the condition and options, probabilities of benefits and risks, a values clarification exercise, and structured guidance in deliberation and communication. To address the decisional need of relocation to the transplant centre the names of the 5 transplant cities (Vancouver, Edmonton, Toronto, Winnipeg, Montreal) were listed in the decision aid to ensure that patients reviewing the decision aid in other cities like Halifax understood that they would have to relocate to one of the transplant centres. Other statements such as, "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 centre while waiting for your new lungs" (Canadian CFDA page 3) were included as a prompt for discussion with the social workers at the referral site. Information about how to cope with anxiety and depression was not included in the decision aid. In Step 1 of the decision aid, "How CF affects me now?" (Canadian CFDA page 2) the patient was provided with a section where they could indicate that they were feeling anxious, scared, angry, depressed or unable to cope similarly acting as a prompt for the CF team to investigate further as needed. Measures for anxiety were not included in the evaluation phase as Bekker et al. (2003) have shown in their systematic review that these measures are insufficient and that administering decision aids do not affect patients' anxiety.

Subsequently, we conducted a prospective evaluation in a randomized controlled trial to determine whether use of the decision aid increased knowledge

about the options, improved realistic expectations, and decreased decisional conflict in adult patients with CF. (Chapter 4) We found that patients randomized to the decision aid had greater knowledge about their options (P <0.0001) and had more realistic expectations about the benefits and risks of lung transplantation (P <0.0001) compared to those randomized to usual care. The total decisional conflict score was significantly lower in the decision aid group three weeks post randomization compared to the usual care group (11.6 vs 20.4, P = 0.0007). We also demonstrated that decisions were durable with 88% of patients in the decision aid group and 75% in the usual care group maintaining the same choice 12 months after randomization (P = 0.06).

This thesis is the first of its kind to describe decisional needs of patients considering referral for lung transplantation for end stage CF. Moreover, a decision aid focused on this issue has been developed and evaluated. Such a tool may be useful in addressing the information needs of patients and may be helpful to minimize practice variations and referral variability seen among CF centers.

Implications for Nursing Practice and the Advanced Practice Nurse

Nurses are well positioned in the CF clinics to support patients who are facing the decision about referral for lung transplantation. The Canadian CF Foundation provides salary support to the 38 CF clinics across Canada to ensure that both children and adults with CF receive specialized care. This is similar throughout the United States, Europe and Australia.

The specialized care supported by the Canadian CF Foundation encourages patients to be seen by the CF team regularly, approximately every three to four months. The CF team includes a physician, registered nurse, dietician, social worker, and a physiotherapist. The nurse performs the initial assessment when a patient visits the clinic including height, weight, vital signs, current medications and a brief history of how the patient is feeling. Most of the nurses have the role of an advanced practice nurse (APN). The domains of their practice include clinical practice, consultation, research, education and leadership (CNA, 2003).

As a practitioner with the CF team, the nurse could assess all patients using the five characteristics of direct clinical care: (Hamric, Spross & Hanson, p. 146, 2005) holistic perspective; formation of partnership with patient and family; expert clinical thinking and skilful performance; use of research evidence to guide one's practice; and use of diverse approaches to health and illness management; by performing a comprehensive assessment and evaluation of every patient with CF who visits the clinic and then following up with them at quarterly visits. During this comprehensive assessment the nurse could talk to the patient, their family and the multidisciplinary team and convey the findings of the assessment and discuss strategies with the staff. The nurse could use evidence based research to guide her suggestions for care. The nurse may advocate a shared decision making approach to screen for decisional conflict and provide decision support including resources and decision tools such as decision aids (Stacey et al., 2008).

To ensure that research based knowledge is put into practice (CNA, 2008) by the nurse, the decision aid can be disseminated and implemented using a similar procedure that was used in the evaluation study (Chapter 4). The patient could receive an education session on the process of referral and the risks and benefits of lung transplantation from their CF physician and then receive a copy of the decision aid or the web address where they can view it on-line. The on-line version produces a summary sheet of the patient responses that they can print and the paper copy has a summary sheet that can be completed by the patient. The patient can bring the summary sheet to their next clinic appointment for follow-up discussion with the CF team.

Implications for Education

Health Care Providers

Locally, the nurse could work with the CF team to become trained in the use of decision aids. The team could plan how the decision aid is included in the clinical care pathways. This can be accomplished by embedding decision support alert cues into these care pathways so that patients with a forced expiratory volume less than 40% would be offered a decision aid and would provide them and their family members ample opportunity to review the information, ask questions to the nurse and CF team, and ultimately make an informed choice. Embedding decision support for cancer services is currently being done at the Ottawa Hospital (O'Connor et al., 2007) and reminders have been shown to be effective in changing practitioners practice (Grimshaw et al., 2004).

Public Education

Patient and family access to information is a pre-requisite for active participation in decision making (Coulter & Ellins, 2007) and nationally, the nurse can work with the Canadian CF Foundation to heighten patient awareness of the shared decision making process that should occur with their CF team when making the decision about referral for lung transplantation as well as all other treatment decisions that they are asked to consider. Current thinking by some surgeons, ethicists, lawyers and policy makers support that the informed consent process should be replaced by a thorough documentation of provision of balanced, evidence-based information on all options (Weinstein, Clay, & Morgan, 2007). This would include using decision aids as an adjunct where they exist with discussion of the benefits and risks of each option and the likelihood that they will occur. Additional components would involve clarification of patient's values and their preferred role in decision making with the treatment decision being arrived at through discussion between the health care providers and patient. Most importantly is that the framing and language of the information being provided must be understandable to the patient (Weinstein, Clay, & Morgan, 2007).

Implications for Policy

The Registered Nurses Association of Ontario (RNAO) continues to solicit funds from the Ontario Ministry of Health and Long Term Care into the Nursing Best Practice Guideline Program to develop tools to help nurses engage in evidence based practice (RNAO, 2009). As there are no best practice guidelines

for patients with CF, an opportunity exists for the nurse to work with professional organizations, patients, and advocacy groups to help create such guidelines and to ensure that the findings from this research are incorporated.

Implications for Future Research

There are several areas to consider for further research. These include subsequent evaluations of the lung transplantation decision aid which would be to evaluate longer term patterns of patients' decision making. Our randomized controlled trial did monitor the durability of the decision for 12 months, but future studies should continue to monitor patients until post transplantation or death.

In addition, although the prospective study did contain an "integrated knowledge translation" approach in that the people who will ultimately use the decision aid (nurse, CF clinician) were meaningfully engaged in the research process (Canadian Institutes of Health Research, 2008) future research needs to be performed to ensure that practitioners are continuing to use the decision aid and that practitioners using the decision aid have the necessary training and coaching experience. A recent study by Stacey et al. found that there is a need to enhance the decision support knowledge and skills of nurses in practice (Stacey, O'Connor, Graham, & Pomey, 2006) and effective interventions including online decision support tutorials are available (O'Connor & Jacobson, 2007) and should be incorporated into future studies.

Other consistent findings from the literature indicate that the transfer of research findings into practice is slow and hap-hazard (Graham et al, 2006); therefore, to ensure that the practitioners are using the decision aid for lung

transplantation our group will pursue a research grant in knowledge translation using the knowledge-to-action strategy proposed by Graham et al. This research will enable us to assess barriers of use among the healthcare professionals, patients, and families and to tailor implementation strategies to address these barriers. The research will be a combination of both qualitative and quantitative designs.

Another important issue for the ongoing use of the lung transplantation decision aid is the need for updating the decision aid based on new evidence (Elwyn et al. 2006). There are several ongoing studies of new medical regimens with CF patients that may become conventional therapies in the future including inhaled denufosol tetrasodium and inhaled L-arginine (Deterding et al, 2007; Grasemann, Kurtz, & Ratjen, 2006; McCoy et al., 2008) that would need to be incorporated into the decision aid. As there is very little information in the literature regarding the risks and adverse effects of transplantation surgery in prospective populations and even less information from patient and caregiver perspectives on the process of decision making it is our hope that the findings of our research study will stimulate others to explore these areas.

Lastly, further research in decision aids could be explored for pediatric CF patients who are being considered for referral for lung transplantation and for their parents. Other possible topics for decision aids include male sterility options (Havasi, Keiles, Hambuch, Sorscher, & Kammesheidt, 2008) and treatments for early menopausal symptoms in females (Johannesson, Ludviiksdottir, & Janson, 2000).

Overall Summary

In conclusion, this study has made explicit the decision making needs of CF patients considering referral for lung transplantation and provided an effective tool for translating evidence on transplant to enhance patient's knowledge and expectations. Furthermore this tool was evaluated in 14 CF programs in Canada and Australia. Subsequent initiatives are required to ensure sustainable use of this tool and nurses are ideally positioned to recognize patients experiencing decisional conflict and facilitating patient involvement in shared decision making.

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Appendix A

When your lung function is getting worse...

Should you be referred for a lung transplant?

A decision aid for adults with cystic fibrosis

This decision aid is for you if you have cystic fibrosis and:

- You are 18 years and older
- Your lung function has been getting worse (less than or equal to 40% of normal)
- You want to think about future options when your lungs don't work enough to keep you alive

What is cystic fibrosis (CF)?

It is a genetic disease that affects many body systems. It causes the body to produce abnormally thick, sticky mucous that is difficult to clear. This mucous traps bacteria and leads to chronic infection that damages the lungs. The mucous makes it difficult for air to move in and out of the lungs and causes shortness of breath. It can also cause problems in the liver, pancreas, and with digestion. In Canada, half of the patients with CF live beyond 37 years of age. Most people with CF die of lung disease.

When CF gets worse...

As time goes by, you may have more frequent chest infections and more trouble with your breathing. The infections cause a decline in your lung function. Generally when your lung function is less than 30% of normal your doctor would consider referring you for lung transplantation. At this time, your expected survival without transplantation is approximately 2-3 years.

We realize that you may be reviewing this material when your lungs are still working well enough and lung transplantation is not something you will need in the near future. However, most patients with CF eventually have to consider this option at some point.

What are your options?

- Not to be referred for lung transplantation.
- To be referred for lung transplantation.

Working through the 5 steps of this decision aid will help you decide.

Step 1: Think about how CF affects you now
Step 2: Think about the options, benefits and risks
Step 3: Choose the role you prefer in decision making
Step 4: Find out what else you need to prepare for decision making
Step 5: Plan the next steps

Step 1: Think about how CF affects you now.

How does CF affect your life? Check ☑ any of these that apply.

Breathing

- ☐ shortness of breath
- ☐ coughing
- ☐ coughing up blood
- ☐ coughing up phlegm
- ☐ frequent chest infections
- ☐ frequent hospitalizations

Daily Activity and Lifestyle

- ☐ cannot work or go to school or reduced working hours
- ☐ difficulty maintaining weight
- ☐ less energy
- ☐ difficulty with daily activities (e.g. bathing, preparing meals)
- ☐ short of breath when walking or exercising
- ☐ increasing fatigue

Emotional

- ☐ feeling anxious
- ☐ feeling scared
- ☐ feeling depressed or unable to cope
- ☐ feeling angry or irritable

Social

- ☐ being unable to participate in social activities with family and friends
- ☐ feeling embarrassed in public because of coughing and sputum production
- ☐ feeling isolated

What are you doing to manage your CF? Check ☑ any of these that apply.

Breathing

- ☐ bronchodilators (e.g. ventolin, serevent or oxeze)
- ☐ inhaled antibiotics (e.g. tobramycin, colistin)
- ☐ anti-inflammatories (e.g. ibuprofen, flovent, pulmicort)
- ☐ mucus-thinning agents (e.g. pulmozyme, hypertonic saline)
- ☐ antibiotics
- ☐ oxygen

Daily Activity and Lifestyle

- ☐ regular exercise
- ☐ nutrition supplements (e.g. shakes, puddings)
- ☐ pancreatic enzyme supplements
- ☐ chest physio
- ☐ tube feeding

Emotional

- ☐ talking about feelings with family, friends and CF team
- ☐ taking things one day at a time
- ☐ praying, seeking spiritual support

Alternative Therapy

- ☐ herbal medicine
- ☐ acupuncture
- ☐ massage therapy
- ☐ chiropractor

Step 2: Think about the options, benefits, and risks.

What are the options?

1. Not to be referred for lung transplantation

- 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 laboured. 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**	○ 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 – 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**	○ 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 centre 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 ICU after surgery is 1-4 days** **Average time in hospital after surgery is 2-4 weeks**	○ Your new lungs will come from a person who has recently died and their 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 – 3 days. You will have tubes in your chest (chest tubes) and lines in your arms (intravenous) and wrist (arterial).
After hospital	**Average time is 3-6 months**	○ You will have to live in or very near your transplant center for several months after your transplant.

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.

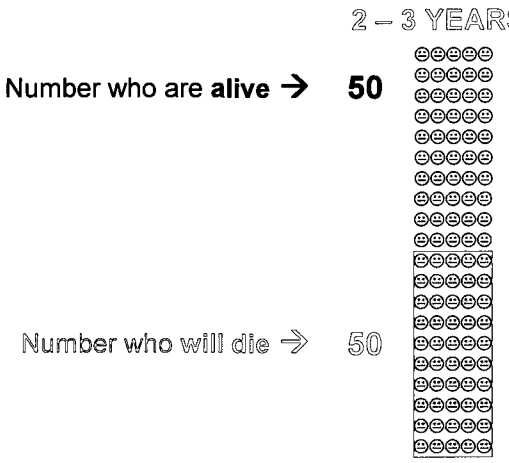
Although your lungs will be healthier, you will still have CF. Lung transplant will not fix other CF health problems like diabetes, digestive problems, osteoporosis or male infertility.

Step 2: Think about the benefits and risks.

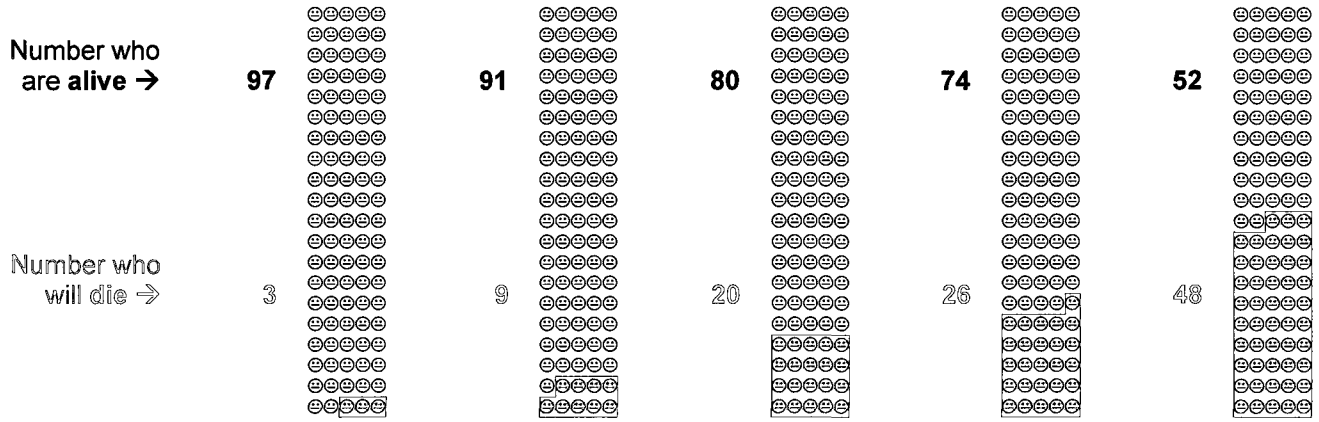
A) What does the research show?

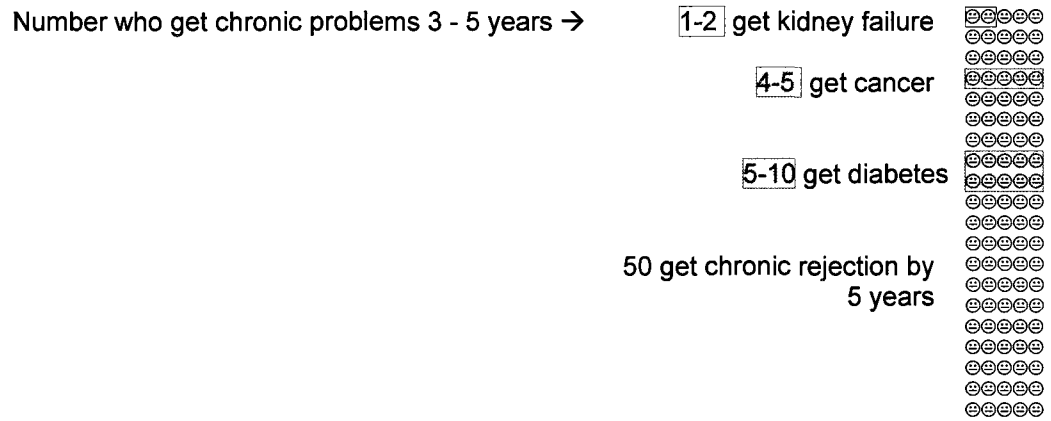
There are no clinical trials comparing options. Study results are based on cases of what happened to someone who had one of the options.
Blocks of 100 faces show the 'best estimate' of what happens to 100 people **with cystic fibrosis who choose different options.** Each face (😐) stands for one person. There is no way of knowing for certain what will happen to you.

Expected Results for Those Who Decline Lung Transplantation [only 1 time period known]



Expected Results After Lung Transplantation





B) What do you think of the benefits and risks of the options?

1. Review the common benefits (reasons to choose) and risks and side effects (reasons to avoid).
2. Add any other reasons that matter to you.
3. Show how much each reason matters to you. Circle one (★) star if it matters a little and up to five (★★★★★) stars if it matters a lot. *Do not circle any stars if it does not matter*

Reasons to Choose

No referral for lung transplant	How much does it matter to you?	Referral for lung transplant	How much does it matter to you?
You avoid the o early risk of death from transplant [3 in 100] and the o long term chance of chronic problems o rejection [50 in 100] o diabetes [5-10 in 100] o cancer [4-5 in 100] o kidney failure [1-2 in 100]	★★★★★	You have a better chance [80 in 100] of surviving for 3 years or longer if you have a transplant compared to no transplant [50 in 100]	★★★★★
You avoid the hassle, stress, and worry of o new care team o extra tests o being on the waitlist o surgery, pain and discomfort in hospital o recovery o taking multiple pills to avoid rejection o possible stay in another city	★★★★★	You are likely to be less short of breath soon after a transplant. You may feel better with new lungs and you may: o breathe easier with less cough o be able to exercise and go back to work or school o have more energy o be able to reach goals and dreams o spend less time on intensive treatment than would be needed if you had your own lungs [oxygen, antibiotics, chest physio, hospital stays]	★★★★★
Other reasons:		Other reasons:	

Which option do you prefer? Check ☑ the option that applies.

☐ Not to be referred for lung transplant ☐ I am unsure ☐ To be referred for lung transplant

Step 3: Choose the role you prefer in decision making.

Check ☑ one.

☐ You prefer to choose on your own after hearing the views of others
☐ You prefer to share the choice with: _____
☐ You prefer that someone else chooses for you, namely: _____

Step 4: Find out what else you need to prepare you for decision making.

Please answer the questions below.
If you answer **'No'** to the questions, discuss them with your doctor.

		Yes	No
Knowledge	Do you know which options are available to you?	☐	☐
	Do you know both the benefits and risks of each option?	☐	☐
Values	Are you clear about which benefits and risks matter most to you?	☐	☐
Support	Do you have enough support and advice from others to make a choice?	☐	☐
	Are you choosing without pressure from others?	☐	☐
Certainty	Do you feel sure about the best choice for you?	☐	☐

Decisional Conflict Scale © A O'Connor 1993, Revised 2004

Find out how well this decision aid helped you learn the key facts.

Check ☑ the best answer. Answers are in Appendix B.

a. Which option has the greatest chance of relieving advanced CF lung symptoms [such as shortness of breath, cough, low energy and poor exercise ability]?
☐ *Lung transplant* ☐ *Not having lung transplant* ☐ *Both are about equal* ☐ *I am unsure*

b. Which option has the greatest chance of chronic complication at 5 years [such as diabetes, cancer, kidney failure]?
☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

c. If 100 people with cystic fibrosis decide not to be referred for lung transplant, about how many will be alive in 2 to 3 years?
☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive
☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ I am unsure

d. If 100 people with cystic fibrosis have lung transplantation, about how many will be alive in 3 years?
☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive
☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ *I am unsure*

© Decision Quality Template, Foundation for Informed Medical Decision Making Question

Step 5: Plan the next steps

List plans, for example: show your balance scale and responses to your doctor and/or family; learn more about the options.

Should you be referred for a lung transplant? (1 page summary)

Step 1: How CF affects me.

Breathing
- ☐ short of breath
- ☐ cough
- ☐ cough blood
- ☐ cough phlegm
- ☐ frequent chest infections
- ☐ frequent hospitalizations

Daily activity
- ☐ cannot work or go to school or reduced working hours
- ☐ difficulty with daily activities
- ☐ difficulty maintaining weight
- ☐ short of breath when walking or exercising
- ☐ less energy
- ☐ increasing fatigue

Emotional
- ☐ feeling anxious
- ☐ feeling depressed or unable to cope
- ☐ feeling scared
- ☐ feeling angry or irritable

Social
- ☐ unable to participate in social activities
- ☐ feeling isolated
- ☐ embarrassed in public because of cough and sputum

- ☐ bronchodilators
- ☐ anti-inflammatories
- ☐ antibiotics
- ☐ *inhaled antibiotics*
- ☐ mucus-thinning agents
- ☐ oxygen
- ☐ regular exercise
- ☐ nutrition supplements
- ☐ pancreatic enzyme supplements
- ☐ chest physio
- ☐ tube feeding

- ☐ herbal medicine
- ☐ acupuncture
- ☐ massage therapy
- ☐ *chiropractor*

- ☐ talking about feelings with family, friends & CF team
- ☐ taking things one day at a time
- ☐ praying, seeking spiritual support

Step 2: My opinion of the options, benefits, and risks.

Reasons to Choose

No referral for lung transplant	How much it matters	Referral for lung transplant	How much it matters
You avoid the early risk of death from transplant [3 in 100] and the long term chance of chronic problems such as rejection [50 in 100]; diabetes [5-10 in 100]; cancer [4-5 in 100]; kidney failure [1-2 in 100]	★★★★★	You have a better chance [80 in 100] of surviving for 3 years or longer if you have a transplant compared to no transplant [50 in 100]	★★★★★
You avoid the hassle, stress, and worry of new care team, extra tests, being on the waitlist, surgery, pain and discomfort in hospital recovery, taking multiple pills to avoid rejection, possible stay in another city	★★★★★	You are likely to be less short of breath soon after a transplant. You may feel better with new lungs and you may: breathe easier with less cough; be able to exercise and go back to work or school; have more energy; be able to reach goals and dreams; spend less time on intensive treatment than would be needed if you had your own lungs	★★★★★
Other reasons:		Other reasons:	

Which option do you prefer?

☐ Not to be referred for lung transplant ☐ I am unsure ☐ To be referred for lung transplant

Step 3: The role you prefer in decision making.

- ☐ You prefer to choose on your own after hearing the views of others
- ☐ You prefer to share the choice with:
- ☐ You prefer that someone else chooses for you, namely:

Step 4: Find out what else you need to prepare you for decision making.

		Yes	No
Knowledge	Do you know which options are available to you?	☐	☐
	Do you know <u>both</u> the benefits and risks of each option?	☐	☐
Values	Are you clear about which benefits and risks <u>matter most</u> to you?	☐	☐
Support	Do you have enough support and advice from others to make a choice?	☐	☐
	Are you choosing without pressure from others?	☐	☐
Certainty	Do you feel sure about the best choice for you?	☐	☐

How well this decision aid helped you learn the key facts.

a. Which option has the <u>greatest</u> chance of relieving advanced CF lung symptoms
☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

b. Which option has the <u>greatest</u> chance of chronic complication at 5 years
☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

c. If 100 people with cystic fibrosis decide not to be referred for lung transplant, about how many will be alive in 2 to 3 years?
☐ between 1 and 10 ☐ between 11 and 40 ☐ between 41 and 60 ☐ between 61 and 100 ☐ I am unsure

d. If 100 people with cystic fibrosis have lung transplantation, about how many will be alive in 3 years?
☐ between 1 and 10 ☐ between 11 and 40 ☐ between 41 and 60 ☐ between 61 and 100 ☐ I am unsure

Step 5: Next steps

Appendix A: Information about the authors

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& The Prince Charles Hospital, Brisbane, Australia
&& Royal Prince Alfred Hospital, Sydney, Australia

Funder: Physician Services Foundation Incorporated and Ontario Thoracic Society

Date: August 2006
Next update due 2008

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Appendix B: Answers to questions in Step 4

a. lung transplant
b. lung transplant
c. between 41-60
d. between 61-100

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Glossary

Kidney Failure. Kidney failure is when your kidneys lose their ability to perform their main function of taking excess fluid and waste material from your blood. Loss of kidney function that develops gradually over time is called chronic kidney failure. Patients who suffer bad kidney failure may need to go on dialysis.

Diabetes. Diabetes or elevated sugar levels may develop after transplantation because of the medications that you are required to take. If diabetes develops after transplant you may need to go onto insulin injections.

Chronic Rejection. Chronic rejection is when your transplanted lungs gradually stop working. This can cause gradual worsening shortness of breath. In extreme cases chronic rejection will lead to death or the need for a second lung transplant.

What it means to answer 'no' to the questions in **Step 4** asking about what else you need. The more **'no'** answers a person has, the more likely they are to delay their decision, change their mind, be dissatisfied with their choice, express regret with the decision they made, and blame their doctors for bad outcomes. Therefore it is important to discuss your needs with your doctor and others so that you answer **'yes'** to most questions.

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This decision aid is being tested to see if it meets the International Patient Decision Aid Standards (IPDAS) Collaboration global standards (http://ipdas.ohri.ca).

Appendix B

Decision Aid Summary Using the IPDAS Criteria from the A-Z Inventory downloaded April 28, 2009

IPDAS Checklist

Content	Answer
1. The decision aid describes the condition (health or other) related to the decision.	**Yes**
2. The decision aid describes the decision that needs to be considered (the index decision).	**Yes**
3. The decision aid lists the options (health care or other).	**Yes**
4. The decision aid describes what happens in the natural course of the condition (health or other) if no action is taken.	**Yes**
5. The decision aid has information about the procedures involved (e.g. what is done before, during, and after the health care option).	**Yes**
6. The decision aid has information about the positive features of the options (e.g. benefits, advantages).	**Yes**
7. The decision aid has information about negative features of the options (e.g. harms, side effects, disadvantages).	**Yes**
8. The information about outcomes of options (positive and negative) includes the chances they may happen.	**Yes**
9. The decision aid has information about what the test is designed to measure.	**NA**
10. The decision aid describes possible next steps based on the test results.	**NA**
11. The decision aid has information about the chances of disease being found with and without screening.	**NA**
12. The decision aid has information about detection and treatment of disease that would never have caused problems if screening had not been done.	**NA**
13. The decision aid presents probabilities using event rates in a defined group of people for a specified time.	**Yes**
14. The decision aid compares probabilities (e.g. chance of a disease, benefit, harm, or side effect) of options using the same denominator.	**Yes**
15. The decision aid compares probabilities of options over the same period of time.	**No**
16. The decision aid uses the same scales in diagrams comparing options.	**Yes**
17. The decision aid asks people to think about which positive and negative features of the options matter most to them.	**Yes**
18. The decision aid makes it possible to compare the positive and negative features of the available options.	**Yes**
19. The decision aid shows the negative and positive features of the options with equal detail.	**Yes**

Development Process	**Answer**
20. Users (people who previously faced the decision) were asked what they need to prepare them to discuss a specific decision.	**Yes**
21. The decision aid was reviewed by people who previously faced the decision who were not involved in its development and field testing.	**Yes**
22. People who were facing the decision field tested the decision aid.	**Yes**
23. Field testing showed that the decision aid was acceptable to users (the general public & practitioners).	**Yes**
24. Field testing showed that people who were undecided felt that the information was presented in a balanced way.	**Yes**
25. The decision aid provides references to scientific evidence used.	**Yes**
26. The decision aid reports the date when it was last updated.	**Yes**
27. The decision aid reports whether authors of the decision aid or their affiliations stand to gain or lose by choices people make after using the decision aid.	**No**
28. The decision aid (or available technical document) reports readability levels.	**No**
Effectiveness	**Answer**
29. There is evidence that the decision aid (or one based on the same template) helps people know about the available options and their features.	**Yes**
30. There is evidence that the decision aid (or one based on the same template) improves the match between the features that matter most to the informed person and the option that is chosen.	**Yes**

Appendix C

Consensus guidelines specific for selection of patients with cystic fibrosis for lung transplantation

Indications

- $FEV_1 \leq 30\%$ of predicted
- Rapid, progressive respiratory deterioration with $FEV_1 > 30\%$ of predicted
  - Increasing hospitalizations
  - Massive hemoptysis
  - Rapidly falling FEV_1
  - Increasing cachexia
- Female gender
- Resting room air blood gas with $PaC0_2 > 50$ mm Hg or $PaO_2 < 55$ mm Hg

Relative contraindications

- Multiple antibiotic resistant *Pseudomonas aeruginosa*
- *Burkholderia cepacia*

Caution

- Pleural fibrosis

(reproduced from Liou TG, Cahill BC, Adler FR, Marshall BC. Selection of patients with cystic fibrosis for lung transplantation. Curr Opin Pul Med 2002;8:535-541)

APPENDIX D

Ottawa Decision Support Framework

Assess Needs (Determinants of Decisions)	Provide Decision Support	Evaluate
Perceptions of Decision Knowledge Expectations Values Decisional conflict Stage of decision making Predisposition **Perceptions of Others** Perceptions of others' opinions & practices Support Pressures Roles in decision making **Resources to Make Decision** Personal Previous experience Self-confidence Motivation Skill in decision making External Support (information, advice, emotional, instrumental, financial, professional help) from social networks and agencies **Characteristics** Client: age, sex, marital status, education, occupation, culture, locale, medical diagnosis & duration, health status Practitioner: age, sex, education, specialty, culture, practice locale, experience, counseling style	**Provide access to information re:** • health situation • options • outcomes • other's opinions and choices **Re-align expectations of outcomes** **Clarify personal values for outcomes** **Provide guidance/coaching in:** • steps in decision making • communicating with others • handling pressure • accessing support & resources	**Decision making** Reduced decisional conflict Improved knowledge Realistic expectations & norms Clear values Agreement between values &choice Implementation of chosen option Satisfaction with decision making **Outcomes of Decision** Persistence with choice Improved quality of life Reduced distress Reduced regret Informed use of resources

APPENDIX E

Definitions of Determinants of Decisions in the Ottawa Decision Support Framework

Participants' Perceptions of the Decision	
Knowledge	cognizance of the health problem or situation, options, and outcomes
Expectations of outcomes	perceived likelihood or probability of outcomes of each option
Values for outcomes	desirability or personal importance of outcomes of options
Decisional conflict	uncertainty about course of action to take
Stage of decision making	phase of decision making in the context of stages of change: not thinking about the options (interested in considering further?); considering the options; close to selecting an option (interested in re-considering options?); taking steps towards implementing option (interested in re-considering options?); have already carried out choice (interested in re-considering options?)
Predisposition	degree to which a person is leaning strongly towards choosing an option or is uncertain
Participants' Perceptions of Others Involved in Decision Making	
Perception of others' opinions and practices	perceptions of what others decide or what others think is the appropriate choice. For the client, important others may include their spouse, family, peers, and practitioner(s). For the practitioner, it may include the client, professional peers, and personal network
Support	informational, emotional, and tangible help from important others to bolster and sustain decision making
Pressure	perception of persuasion, influence, coercion from important others to select one alternative
Role in decision making	the way a participant is or wants to be involved in decision making with others; do they wish to make the choice themselves after considering other opinions, do they want to share decision making with someone else, do they want others to make the decision after considering their opinion
Participants' Resources for Decision Making	
Personal	
Previous Experience	previous exposure to the situation, options, outcomes, decision making process
Self-confidence	belief in one's abilities in decision making, including shared decision making
Motivation	readiness and interest in decision making, including shared decision making
Skill	abilities in making and implementing a decision
External	assets from others that are required to make and implement the decision
Type	availability and access to information, advice, emotional support, instrumental help, financial assistance, and health and social services
Source	social networks, professional networks, support groups, voluntary agencies, and the formal health care, education, and social sectors
Participants' Characteristics	
Client/Patient	age, gender, education, marital status, ethnicity, occupation, locale, diagnosis and duration of condition, health status (physical, emotional, cognitive, social
Practitioner	age, gender, ethnicity, clinical education and specialty, practice locale, years of experience, counseling style

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Appendix F

PARTICIPANT INFORMATION AND CONSENT FORM

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

Principal Investigators: K. Vandemheen, BScN, Drs Shawn Aaron and Annette O'Connor

Purpose of the Study:
Researchers at the Ottawa Hospital are studying the best way to inform cystic fibrosis patients about being referred for bilateral lung transplantation (replacement of both lungs). This research will help us prepare better educational materials that can be used in clinical practice, so patients can make informed choices. You will be assigned to one of two learning strategies by a random method (similar to tossing a coin). You will be asked to review the educational material and answer some questionnaires about the information presented.

Description of the Study:
We are asking you to:
1. Complete a questionnaire on your opinions regarding being referred for lung transplantation, which should take 10-20 minutes to complete.

2. Meet with your CF physician to discuss the option of lung transplantation. This will take 10-15 minutes.

3. Review the learning strategy you are assigned to. This will take 30 to 45 minutes.

4. Respond to a phone interview three weeks following your visit to discuss what you have learned, your decision about therapy, and how difficult it was to make this decision. The interview will take 10- 20 minutes.

5. Respond to 1 other interview by phone at 12 months to discuss such things as whether your decision changed and how you feel about your decision. This will take 10 minutes.

Potential Benefits:
By participating in this study, you may learn more about being referred for lung transplantation and the available treatments.

Potential Risks:
Participation in this study will require you to think about the future when you may have to make a decision about whether to be referred for a lung transplant. Some people may find thinking about lung transplantation upsetting.

Study Costs:
You will be provided with $20 to cover the cost of parking.

Confidentiality
All information you provide will be kept confidential. Your data will only be identified by a unique ID number. Only the study team and study nurse will have access to the confidential data. If the data is published, your identity will be not be revealed. No records bearing your name will leave the Ottawa Hospital.

Voluntary Participation/Withdrawal
Your participation in the study is voluntary. You can withdraw your consent and stop your participation in the study at any time and for any reason. If you choose not to participate your decision will have no impact on your care now, or in the future.

If you have any questions about the study, now or later, you may call the study coordinator at 613-737-8259. If you have any questions about your rights as a research subject, you may contact the Chairperson of the Ottawa Hospital Research Ethics Board at 613-798-5555 x14902.

Consent
I will be given a copy of this consent form for my records.

With full knowledge of this, I voluntarily consent to participate in the study.

_____
Patient's Name (printed)

_____ _____
Patient's Signature Date

_____
Investigator/Delegate Name (printed)

_____ _____
Investigator/Delegate Signature Date

(valid until December 13, 2008)

Université d'Ottawa University of Ottawa
Service de subventions de recherche et déontologie Research Grants and Ethics Services

HEALTH SCIENCES AND SCIENCE RESEARCH ETHICS BOARD

CERTIFICATION OF ETHICS APPROVAL

This is to certify that the University of Ottawa Health Sciences and Science Research Ethics Board (REB) examined the application for extension of ethics approval for the research project **Development of a Decision Aid for Adult Cystic Fibrosis Patients Considering Bilateral Lung Transplantation (H 05-07-05)** submitted by Professor Annette O'Connor of the School of Nursing at the University of Ottawa and Ms. Katherine Vandemheen at the Ottawa General Hospital.
This project received initial ethics approval on October 23, 2007 by the REB as meeting appropriate ethical standards set out in the Tri-Council Policy Statement and in the Procedures of the University of Ottawa Research Ethics Boards. The University of Ottawa REB members accordingly gave it a one-year extension of ethics approval. This ethics renewal certification is valid until October 23, 2009.

Germain Zongo
Protocol Officer for Ethics in Research
For Dr. Daniel Lagarec, Chair of the
Health Sciences and Science REB

October 7, 2008
Date

550, rue Cumberland 550 Cumberland Street
Ottawa (Ontario) K1N 6N5 Canada Ottawa, Ontario K1N 6N5 Canada

(613) 562-5841 • Téléc./Fax (613) 562-5338
http://www.uottawa.ca/services/research/rge/index.html

Appendix I
A Decision Aid for Adult cystic Fibrosis Patients Considering Referral for Lung Transplantation

Patient Questionnaire

1. How would you rate the amount of information in the decision aid? Please check ☑ one.
   - ☐ Much less than I needed
   - ☐ A little less than I needed
   - ☐ About the right amount of information
   - ☐ A little more information than I needed
   - ☐ A lot more information than I needed

2. Did you find information in the decision aid upsetting? Please check ☑ one.
   - ☐ Not at all upsetting
   - ☐ A little upsetting
   - ☐ Somewhat upsetting
   - ☐ Very upsetting

3. How would you rate the length of the decision aid? Please check ☑ one.
   - ☐ Much too long
   - ☐ A little too long
   - ☐ Just about right
   - ☐ Should have been a bit longer
   - ☐ Should have been much longer

4. How balanced was the information about referral for lung transplantation versus no referral for lung transplantation? Please check ☑ one.
   - ☐ Clearly slanted towards referral for lung transplantation
   - ☐ A little slanted towards referral for lung transplantation
   - ☐ Completely balanced
   - ☐ A little slanted towards no referral for lung transplantation
   - ☐ Clearly slanted towards no referral for lung transplantation

5. Did the decision aid present one option as the best overall choice? Please check ☑ one.
   - ☐ No, the decision aid was neutral and balanced
   - ☐ Yes, the decision aid favoured referral for lung transplantation
   - ☐ Yes, the decision aid favoured no referral for lung transplantation

6. How clear was the information in the decision aid? Please check ☑ one.
   - ☐ Everything was clear
   - ☐ Most things were clear
   - ☐ Some things were clear
   - ☐ Many things were unclear

7. How helpful would the decision aid be in helping other people who are facing the same decision? Please check ☑ one.
   - ☐ Very helpful
   - ☐ Somewhat helpful
   - ☐ A little helpful
   - ☐ Not helpful

8. Would you recommend this decision aid to other people who are facing the same decision? Please check ☑ one.
   - ☐ I would definitely recommend it
   - ☐ I would probably recommend it
   - ☐ I would probably not recommend it
   - ☐ I would definitely not recommend it

Please answer the next two questions as though *you had been given the decision aid before making your choice about whether or not to be referred for lung transplantation.*

9. Would this decision aid have . . .

	Not at all	A little	Somewhat	Quite a bit	A great deal
Helped you recognize that a decision needed to be made?	☐	☐	☐	☐	☐
Prepared you to make a better decision?	☐	☐	☐	☐	☐
Helped you think about the benefits and risks of each option?	☐	☐	☐	☐	☐
Helped you think about which benefits and risks are most important?	☐	☐	☐	☐	☐
Helped you know that your values (what matters most) affect the decision?	☐	☐	☐	☐	☐
Helped you organize your own thoughts about the decision?	☐	☐	☐	☐	☐
Helped you think about how involved you wanted to be in this decision?	☐	☐	☐	☐	☐

Preparation for Decision Making Scale (Graham, O'Connor, 1995, revised 2005)

10. Would this decision aid have . . .

	Not at all	A little	Somewhat	Quite a bit	A great deal
Helped you identify the questions you wanted to ask?	☐	☐	☐	☐	☐
Prepared you to communicate your opinion to your doctor?	☐	☐	☐	☐	☐
Prepared you for a follow-up visit with your doctor?	☐	☐	☐	☐	☐
Made a follow-up visit with your doctor run more smoothly?	☐	☐	☐	☐	☐

Preparation for Decision Making Scale (Graham, O'Connor, 1995, revised 2005)

PLEASE WRITE COMMENTS ABOUT THE DECISION AID HERE:

About you

11. What is your age? _____ years

12. What is your gender? Please check ☑ one.
    - ☐ Male
    - ☐ Female

13. What is the highest grade you completed in school? Please check ☑ one.
    - ☐ Some high school
    - ☐ Graduated high school
    - ☐ Some community college or university
    - ☐ Graduated community college or university

Appendix J
A Decision Aid for Adult cystic Fibrosis Patients Considering Referral for Lung Transplantation

Healthcare Provider Questionnaire

Your answers to the next set of questions will help us improve the decision aid.

1. How would you rate the amount of information in the decision aid? Please check ☑ one.
   - ☐ Much less than is needed
   - ☐ A little less than is needed
   - ☐ About the right amount of information
   - ☐ A little more information than is needed
   - ☐ A lot more information than is needed

   Comments:_____

2. Do you find information in the decision aid upsetting for patients? Please check ☑ one.
   - ☐ Not at all upsetting
   - ☐ A little upsetting
   - ☐ Somewhat upsetting
   - ☐ Very upsetting

   Comments:_____

3. How would you rate the length of the decision aid? Please check ☑ one.
   - ☐ Much too long
   - ☐ A little too long
   - ☐ Just about right
   - ☐ Should have been a bit longer
   - ☐ Should have been much longer

   Comments:_____

4. How balanced was the information about referral for lung transplantation versus no referral for lung transplantation? Please check ☑ one.
   - ☐ Clearly slanted towards referral for lung transplantation
   - ☐ A little slanted towards referral for lung transplantation
   - ☐ Completely balanced
   - ☐ A little slanted towards no referral for lung transplantation
   - ☐ Clearly slanted towards no referral for lung transplantation

   Comments:_____

5. Does the decision aid present one option as the best overall choice? Please check ☑ one.
   - ☐ No, the decision aid was neutral and balanced
   - ☐ Yes, the decision aid favoured referral for lung transplantation
   - ☐ Yes, the decision aid favoured no referral for lung transplantation

   Comments:_____

6. How clear is the information in the decision aid? Please check ☑ one.
   - ☐ Everything was clear
   - ☐ Most things were clear
   - ☐ Some things were clear
   - ☐ Many things were unclear

   Comments:_____

7. How helpful would the decision aid be in preparing patients for decision making decision about referral for lung transplantation? Please check ☑ one.
   - ☐ Very helpful
   - ☐ Somewhat helpful
   - ☐ A little helpful
   - ☐ Not helpful

   Comments:_____

8. Would you recommend this decision aid to patients? Please check ☑ one.
   - ☐ I would definitely recommend it
   - ☐ I would probably recommend it
   - ☐ I would probably not recommend it
   - ☐ I would definitely not recommend it

   Comments:_____

9. What suggestions do you have to improve the decision aid?

_____

_____

_____

_____

_____

_____

About You

Professional Training:

- ☐ CF physician
- ☐ CF Clinic RN
- ☐ Transplant Surgeon
- ☐ Transplant RN
- ☐ Respirologist
- ☐ Other (Specify) _____

About how many patients with cystic fibrosis did you discuss lung transplantation with last year?

- ☐ Fewer than 5
- ☐ 5 to 10
- ☐ More than 10

Appendix K

Subject Initials ☐☐☐ Subject Number ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

SUBJECT ELIGIBILITY

Case Report Form

For Study Centre:

Signature of Research Personnel Completing Assessment: _____

Date of Signature: _____

For Coordinating Centre:

Initials and Date of Reviewer: _____

Initials of Data Entry Clerk: _____

Appendix K

Subject Initials ☐☐☐ **Subject Number** ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

SUBJECT ELIGIBILITY

Date Subject Screened: ☐☐☐☐-☐☐-☐☐ (yyyy-mm-dd)

INCLUSION CRITERIA (all questions must be answered "YES")

1. Is the subject ≥ 18 years of age? ☐ No ☐ Yes
2. Does the subject have a confirmed diagnosis of cystic fibrosis? ☐ No ☐ Yes
   (a sweat chloride value higher than 60 mmol/litre or 2 disease-causing mutations)
3. Does the subject have a FEV1 ≤ 40% of predicted? ☐ No ☐ Yes
4. Is the subject willing and able to give informed consent? ☐ No ☐ Yes

EXCLUSION CRITERIA (all questions must be answered "No")

1. Is the subject on a transplant waiting list? ☐ No ☐ Yes
2. Is the subject a lung transplant recipient? ☐ No ☐ Yes
3. Does the CF clinician feel that the subject is too sick for transplant? ☐ No ☐ Yes
4. Has the subject's spouse/significant other or sibling been recruited? ☐ No ☐ Yes
   ***please note only one subject per family will be recruited

If all questions are answered "YES" for the Inclusion Criteria and all questions are answered "NO" for the Exclusion Criteria the subject may be randomized to the study.

Will this subject be randomized to the study? ☐ No ☐ Yes

If 'no', why not? ☐ N/A (reason given above) _____

Appendix K

Subject Initials ☐☐☐ Subject Number ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

VISIT 1 - DAY 0

Case Report Form

For Study Centre:

Signature of Research Personnel Completing Assessment: _____

Date of Signature: _____

For Coordinating Centre:

Initials and Date of Reviewer: _____

Initials of Data Entry Clerk: _____

Subject Initials ☐☐☐ **Subject Number** ☐☐–☐–☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

VISIT 1 - DAY 0

Date Consent Signed: ☐☐☐☐-☐☐-☐☐ (yyyy-mm-dd)

DEMOGRAPHICS

Date of Enrollment: ☐☐☐☐-☐☐-☐☐ (yyyy-mm-dd)

Date of Birth: ☐☐☐☐-☐☐-☐☐ (yyyy-mm-dd) or **Age:** ☐☐

Gender: ☐ Male ☐ Female

Height: ☐☐☐.☐ cm

Weight: ☐☐☐.☐ kg

CURRENT MEDICATIONS *(only include medications taken in the past 3 months)*

Does the patient take inhaled tobramycin or TOBI? ☐ No ☐ Yes

Does the patient take inhaled pulmozyme (DNASe)? ☐ No ☐ Yes

Does the patient take inhaled colisten? ☐ No ☐ Yes

Does the patient take chronic azithromycin? ☐ No ☐ Yes

Subject Initials ☐☐☐ Subject Number ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

VISIT 1 - DAY 0

PATIENT ASSESSMENTS

CONCURRENT MEDICAL CONDITION

Does the patient have a history of any of the following medical conditions?

CF- related diabetes	☐ No	☐ Yes
Pancreatic insufficiency	☐ No	☐ Yes
CF- related liver disease	☐ No	☐ Yes

If **'other'** condition, please specify: _____

PATIENT ASSESSMENTS *(from most recent clinic visit)*

Height: ☐☐☐.☐ cm

Weight: ☐☐☐.☐ kg

Pulmonary Function Results from Pre-Bronchodilator: ***(from most recent clinic visit)***

FEV_1 (L) (measured) ☐.☐☐

FEV_1 (L) % (predicted) ☐☐☐

FVC (L) (measured) ☐.☐☐

FVC (L) % (predicted) ☐☐☐

Appendix K

Subject Initials ☐☐☐ Subject Number ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

VISIT 1 - DAY 0

CLINICAL INFORMATION FOR THE PAST 12 MONTHS

1. **Number of admissions to hospital in past year for any reason:** ☐☐

2. **Number of days spent in hospital in past year for any reason:** ☐☐☐

3. **Number of pulmonary exacerbations in past year:** ☐☐

4a) Number of pulmonary exacerbations in past year requiring admission to hospital: ☐☐

4b) Number of pulmonary exacerbations in past year treated with HOME IV antibiotics: ☐☐

4c) Number of pulmonary exacerbations in past year treated exclusively as outpatients with ORAL antibiotics: ☐☐ (4b. count only those that did not require hospital admission; 4c. count only those that did not require IV antibiotics or hospital admission) The sum of 4a +4b +4c =3)

5. **In the past 12 months, which of the following organisms have been identified in the patients' sputum culture.**

☐ Burkholderia cepacia ☐ Pseudomonas aeruginosa

☐ Stenotrophomonas Maltophilia ☐ Staphylococcus aureus

☐ Alcaligenes ☐ Others:_____

Subject Initials ☐☐☐ Subject Number ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

VISIT 1 - DAY 0

What is the highest grade you completed in school? Please check ☑ one.

- ☐ 8th grade or less
- ☐ Some high school
- ☐ Graduated high school
- ☐ Some college
- ☐ Graduated college
- ☐ Graduated university

How far along are you with the decision to be referred for lung transplantation? Please check ☑ one.

- ☐ I have not yet thought about options
- ☐ I am considering the options
- ☐ I am close to choosing an option
- ☐ I have already chosen an option

At this time, which option are you leaning toward? Please check ☑ one.

- ☐ To be referred for lung transplantation
- ☐ Not to be referred for lung transplantation
- ☐ Unsure

What do you think about the options?

	Yes	Probably Yes	Unsure	Probably no	No
Do you know which options are available to you?	☐	☐	☐	☐	☐
Do you know the benefits of each option?	☐	☐	☐	☐	☐
Do you know the risks and side effects of each option?	☐	☐	☐	☐	☐
Are you clear about which benefits matter most to you?	☐	☐	☐	☐	☐
Are you clear about which risks and side effects matter most to you?	☐	☐	☐	☐	☐
Are you clear about which is more important to you (the benefits or the risks and side effects)?	☐	☐	☐	☐	☐

Subject Initials ☐☐☐ Subject Number ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

VISIT 1 - DAY 0

How do you feel about making a decision?

	Yes	Probably Yes	Unsure	Probably no	No
Do you have enough support from others to make a choice?	☐	☐	☐	☐	☐
Are you choosing without pressure from others?	☐	☐	☐	☐	☐
Do you have enough advice to make a choice?	☐	☐	☐	☐	☐
Are you clear about the best choice for you?	☐	☐	☐	☐	☐
Do you feel sure about what to choose?	☐	☐	☐	☐	☐
Is this decision easy for you to make?	☐	☐	☐	☐	☐
Do you feel you have made an informed choice?	☐	☐	☐	☐	☐
Does your decision show what is important to you?	☐	☐	☐	☐	☐
Do you expect to stick with your decision?	☐	☐	☐	☐	☐
Are you satisfied with your decision?	☐	☐	☐	☐	☐

We would like to know how familiar you are with the options before you review the information that we have prepared for you.

Which option has the <u>greatest</u> chance of relieving advanced CF lung symptoms [such as shortness of breath, cough, low energy and poor exercise ability]?

☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

Which option has the <u>greatest</u> chance of chronic complication at 5 years [such as diabetes, cancer, kidney failure]?

☐ Lung transplant ☐ Not having lung transplant ☐ Both are about equal ☐ I am unsure

If 100 people with cystic fibrosis decide not to be referred for lung transplant, about how many will be alive at 2 to 3 years?

☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive
☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ I am unsure

If 100 people with cystic fibrosis have lung transplantation, about how many will be alive at 3 years?

☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive

Subject Initials ☐☐☐ **Subject Number** ☐☐–☐–☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

VISIT 1 - DAY 0

☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ I am unsure

Your answers to the next set of questions will help us to know how strongly you feel about the choices you face.

On a scale from 1 to 10, where 1 is not at all important and 10 is very important	**Not at all Important**									**Very Important**
1. How important is it for you to live as long as possible?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10
2. How important is it to you to avoid complications of lung transplantation such as early risk of death, rejection, diabetes, cancer, kidney failure?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10
3. How important is it for you to be able to improve your breathing, exercise, energy and time spent on CF treatment?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10
4. How important is it for you to avoid the hassle, stress and worry of transplantation such as being on a transplant waiting list, possibly moving to a new city, having a new care team, having pain and discomfort from surgery?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10

Appendix L

Subject Initials ☐☐☐ **Subject Number** ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 3 WEEKS

Case Report Form

For Study Centre:

Signature of Research Personnel Completing Assessment: _____

Date of Signature: _____

For Coordinating Centre:

Initials and Date of Reviewer: _____

Initials of Data Entry Clerk: _____

Appendix L

Subject Initials ☐☐☐ **Subject Number** ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 3 WEEKS

Date of 3 week Follow-up: ☐☐☐☐-☐☐-☐☐ (yyyy-mm-dd)

1. About three weeks ago you completed some questionnaires with the study team. Since that time did you call or meet with your doctor to discuss options for lung transplantation? Please check ☑ one.

☐ Yes
☐ No

2. How far along are you with the decision to be referred for lung transplantation? Please check ☑ one.

☐ I have not yet thought about options
☐ I am considering the options
☐ I am close to choosing an option
☐ I have already chosen an option

3. At this time, which option are you leaning toward? Please check ☑ one.

☐ To be referred for lung transplantation
☐ Not to be referred for lung transplantation
☐ Unsure

4. What do you think about the options?

	Yes	Probably Yes	Unsure	Probably no	No
Do you know which options are available to you?	☐	☐	☐	☐	☐
Do you know the benefits of each option?	☐	☐	☐	☐	☐
Do you know the risks and side effects of each option?	☐	☐	☐	☐	☐
Are you clear about which benefits matter most to you?	☐	☐	☐	☐	☐
Are you clear about which risks and side effects matter most to you?	☐	☐	☐	☐	☐
Are you clear about which is more important to you (the benefits or the risks and side effects)?	☐	☐	☐	☐	☐

Appendix L

Subject Initials ☐☐☐ **Subject Number** ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 3 WEEKS

5. How do you feel about making a decision?

	Yes	Probably Yes	Unsure	Probably no	No
Do you have enough support from others to make a choice?	☐	☐	☐	☐	☐
Are you choosing without pressure from others?	☐	☐	☐	☐	☐
Do you have enough advice to make a choice?	☐	☐	☐	☐	☐
Are you clear about the best choice for you?	☐	☐	☐	☐	☐
Do you feel sure about what to choose?	☐	☐	☐	☐	☐
Is this decision easy for you to make?	☐	☐	☐	☐	☐
Do you feel you have made an informed choice?	☐	☐	☐	☐	☐
Does your decision show what is important to you?	☐	☐	☐	☐	☐
Do you expect to stick with your decision?	☐	☐	☐	☐	☐
Are you satisfied with your decision?	☐	☐	☐	☐	☐

6. Did the information materials you were given about the decision ...

	Not at all	A little	Somewhat	Quite a bit	A great deal
Help you recognize that a decision needs to be made?	☐	☐	☐	☐	☐
Prepare you to make a better decision?	☐	☐	☐	☐	☐
Help you think about the benefits and risks of each option?	☐	☐	☐	☐	☐
Help you think about which benefits and risks are most important?	☐	☐	☐	☐	☐
Help you know that the decision depends on what matters most to you?	☐	☐	☐	☐	☐
Help you organize your own thoughts about the decision?	☐	☐	☐	☐	☐
Help you think about how involved you want to be in this decision?	☐	☐	☐	☐	☐

7. Will the information materials you were given about the decision...

	Not at all	A little	Somewhat	Quite a bit	A great deal
Help you identify the questions you want to ask your doctor?	☐	☐	☐	☐	☐
Prepare you to talk to your doctor about what matters most to you?	☐	☐	☐	☐	☐
Prepare you for a follow-up visit with your doctor?	☐	☐	☐	☐	☐

Appendix L

Subject Initials ☐☐☐ **Subject Number** ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 3 WEEKS

8. Who do you think should make the decision? Please check ☑ one.

☐ Mainly my doctor should make the decision, while knowing my opinion.
☐ My doctor and I should both make the decision.
☐ Mainly I should make the decision, while knowing my doctor's opinion

9. We would like to know how familiar you are with the options now that you have reviewed the information we prepared for you.

Which option has the greatest chance of relieving advanced CF lung symptoms [such as shortness of breath, cough, low energy and poor exercise ability]?

☐ Lung transplant
☐ Not having lung transplant
☐ Both are about equal
☐ I am unsure

Which option has the greatest chance of chronic complication at 5 years [such as diabetes, cancer, kidney failure]?

☐ Lung transplant
☐ Not having lung transplant
☐ Both are about equal
☐ I am unsure

If 100 people with cystic fibrosis decide not to be referred for lung transplant, about how many will be alive at 2 to 3 years?

☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive
☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ I am unsure

If 100 people with cystic fibrosis have lung transplantation, about how many will be alive at 3 years?

☐ between 1 and 10 people will be alive
☐ between 11 and 40 people will be alive
☐ between 41 and 60 people will be alive
☐ between 61 and 100 people will be alive
☐ I am unsure

Appendix L

Subject Initials ☐☐☐ **Subject Number** ☐☐–☐–☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 3 WEEKS

10. Your answers to the next set of questions will help us to know how strongly you feel about the choices you face.

On a scale from 1 to 10, where 1 is not at all important and 10 is very important	**Not at all Important**									**Very Important**
How important is it for you to live as long as possible?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10
How important is it to you to avoid complications of lung transplantation such as early risk of death, rejection, diabetes, cancer, kidney failure?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10
How important is it for you to improve your breathing, exercise, energy and time spent on CF treatment?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10
How important is it for you to avoid the hassle, stress and worry of transplantation such as being on a transplant waiting list, possibly moving to a new city, having a new care team, having pain and discomfort from surgery?	☐ 1	☐ 2	☐ 3	☐ 4	☐ 5	☐ 6	☐ 7	☐ 8	☐ 9	☐ 10

Appendix L

Subject Initials ☐☐☐ **Subject Number** ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 3 WEEKS

11. YOUR ANSWERS TO THE NEXT SET OF QUESTIONS WILL HELP US TO IMPROVE THE INFORMATION MATERIALS. (Please check ☑ one for each question)

Did you read all of the information material you were provided?

- ☐ Yes, I read all of the information
- ☐ I read some of the information
- ☐ No, I did not read any of the information

How long did you spend reading and thinking about the information?

- ☐ Less than 1 hour
- ☐ Between 1 hour and 6 hours
- ☐ More than 6 hours

How would you rate the amount of information in the materials you were provided?

- ☐ Much less than I needed
- ☐ A little less than I needed
- ☐ About the right amount of information
- ☐ A little more information than I needed
- ☐ A lot more information than I needed

How balanced was the information in the material you were provided?

- ☐ Clearly slanted towards referral and lung transplantation
- ☐ A little slanted towards referral and lung transplantation
- ☐ Completely balanced
- ☐ A little slanted towards no referral for lung transplantation
- ☐ Clearly slanted towards no referral for lung transplantation

How clear was the information in the material you were provided?

- ☐ Everything was clear
- ☐ Most things were clear
- ☐ Some things were clear
- ☐ Many things were unclear

How helpful was the information material in helping you make a decision about options?

- ☐ Very helpful
- ☐ Somewhat helpful
- ☐ A little helpful
- ☐ Not helpful

Would you recommend these materials to other people who are facing the same decision?

- ☐ I would definitely recommend it
- ☐ I would probably recommend it
- ☐ I would probably not recommend it
- ☐ I would definitely not recommend it

Appendix M

Subject Initials ☐☐☐ Subject Number ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 12 MONTHS

Case Report Form

For Study Centre:

Signature of Research Personnel Completing Assessment: _____

Date of Signature: _____

For Coordinating Centre:

Initials and Date of Reviewer: _____

Initials of Data Entry Clerk: _____

Appendix M

Subject Initials ☐☐☐ **Subject Number** ☐☐-☐-☐☐☐

A Randomized Controlled Trial Evaluating the Impact of Learning Strategies on the Decision Making of Cystic Fibrosis Patients

TELEPHONE FOLLOW-UP IN 12 MONTHS

Date of 12 month follow-up: ☐☐☐☐-☐☐-☐☐ (yyyy-mm-dd)

This first question will be completed by the research staff using the response from page 1, question 3 of the 3 week follow-up.

About a year ago you completed some questionnaires with the CF study team about being referred for lung transplantation.
At that time, you were leaning toward the option of ...

- ☐ Referral for lung transplantation
- ☐ Not to be referred for lung transplantation
- ☐ You were unsure which option you would choose

In the past year, have you discussed these options any further with the CF doctor and/or the clinic team?
Please check ☑ one.
☐ Yes
☐ No

At this time, which option are you leaning toward?
Please check ☑ one.
☐ To be referred for lung transplantation
☐ Not to be referred for lung transplantation
☐ Unsure

In the past year have you been formally referred to a lung transplant program for assessment?
Please check ☑ one.
☐ Yes
☐ No

If 'yes', are you currently on a waiting list for lung transplant?
☐ Yes
☐ No

Have you had a lung transplant in the last year?
Please check ☑ one.
☐ Yes
☐ No

If 'yes', when did you have lung transplant surgery?

☐☐☐☐-☐☐-☐☐ (yyyy-mm-dd)

Cystic Fibrosis and Lung Transplantation

Canadian Cystic Fibrosis Foundation

Margaret Benson, who had a double-lung transplant in 1999, celebrates a gold-medal victory at the World Transplant Games.

The Canadian Cystic Fibrosis Foundation

The Canadian Cystic Fibrosis Foundation (CCFF) is a Canada-wide health charity, with more than 50 volunteer chapters. The Foundation's mandate is to help individuals with cystic fibrosis (CF) principally by funding CF research and supporting high quality clinical and transplant care. The Foundation also provides educational materials for the CF community and the general public, and raises funds to support its programs. The Foundation's goal is to ensure that every dollar raised works for all persons with CF in Canada, without regard to province or community of residence. This philosophy has been paying off for more than 45 years. In 1960, children diagnosed with CF weren't expected to live long enough to attend kindergarten. Now, half of all Canadians who have CF are living into their late 30s and beyond, and almost half of all Canadians with CF are over the age of 18.

Introduction

Transplantation is a surgical procedure that replaces severely damaged organs with healthy organs. When medical management alone can no longer maintain a person's health or organ function, improvements in the field of organ transplantation have opened the door to this life-extending treatment. While transplantation is a procedure that offers new hope, it also presents new challenges and responsibilities.

Individuals with cystic fibrosis in need of a transplant usually require new lungs, but may also need liver, kidney, or heart transplants. This brochure focuses on lung transplantation, and is written for individuals with cystic fibrosis who are contemplating or waiting for a lung transplant. This brochure also provides information for their families and friends. Please note that programs offered at individual transplant centres may differ slightly, and each centre will provide details specific to its program.

Despite the ongoing progress being made in CF treatment, cystic fibrosis remains fatal and most CF deaths are caused by lung disease. For many individuals with CF, years of chronic lung infections damage and destroy lung tissue. In turn, this damage leads to decreased lung function and the need for new lungs.

To qualify for a lung transplant and to be added to a transplant waiting list, individuals with CF need to go through a referral and assessment process. Prior to arriving at a stage of health when a transplant could be critical to survival, it is advisable to be informed about transplantation. Being informed allows individuals with CF and their family members the opportunity to learn and ask questions about transplantation. Then, if a transplant is needed, candidates are better equipped to make an informed decision about whether to proceed with the surgery.

Post-transplant, most individuals with CF report improved strength, energy and exercise capacity, and freedom from symptoms such as constant coughing, and shortness of breath. Ongoing advances in transplantation procedures and care have significantly improved post-transplant outcomes. While transplantation is an important treatment option for damaged CF lungs, it is not a cure. After individuals with CF undergo a lung transplant, cystic fibrosis will still be present in other body cells, such as the cells of the pancreas. However, lungs that are transplanted into recipients' bodies do not have CF because they have the donor's genetic features. Post-transplant, lung recipients will still need to visit either a CF clinic or a transplant centre to help manage other CF-related health issues.

The Transplant Process

Considering candidates for lung transplantation

Cystic fibrosis physicians follow general guidelines to determine if individuals with CF should be considered for transplantation. The main indicator for transplantation is the progression of CF with deterioration of lung function. Other indicators that reveal the need for a transplant include an inability to maintain weight and frequent hospital admissions. It is important to note that to qualify for transplantation, individuals with CF must be ill enough to need a transplant, but well enough to endure the surgery. Some of the test results that are considered by the CF clinic team, when referring a CF patient to a transplant centre, are:

Forced Expiratory Volume (FEV_1):
This test measures the amount of air that can be forcefully blown out of the lungs during the first second of expiration. Individuals may be considered for transplantation if their FEV_1 falls below 30 percent or if there is a sudden, rapid decline in FEV_1.

Hypoxemia < 55 mmHg:
This test measures the extent of lung damage and the lungs' inability to provide enough oxygen for the body to function.

Hypercapnia >45 mmHg:
This test measures the lungs' inability to get rid of carbon dioxide.

Life-threatening events, such as repeated episodes of hemoptysis (coughing up blood from the lungs), frequent infections and/or pneumothoraces (a collection of air between the outside surface of the lungs and the inside surface of the chest wall) may call for earlier referral.

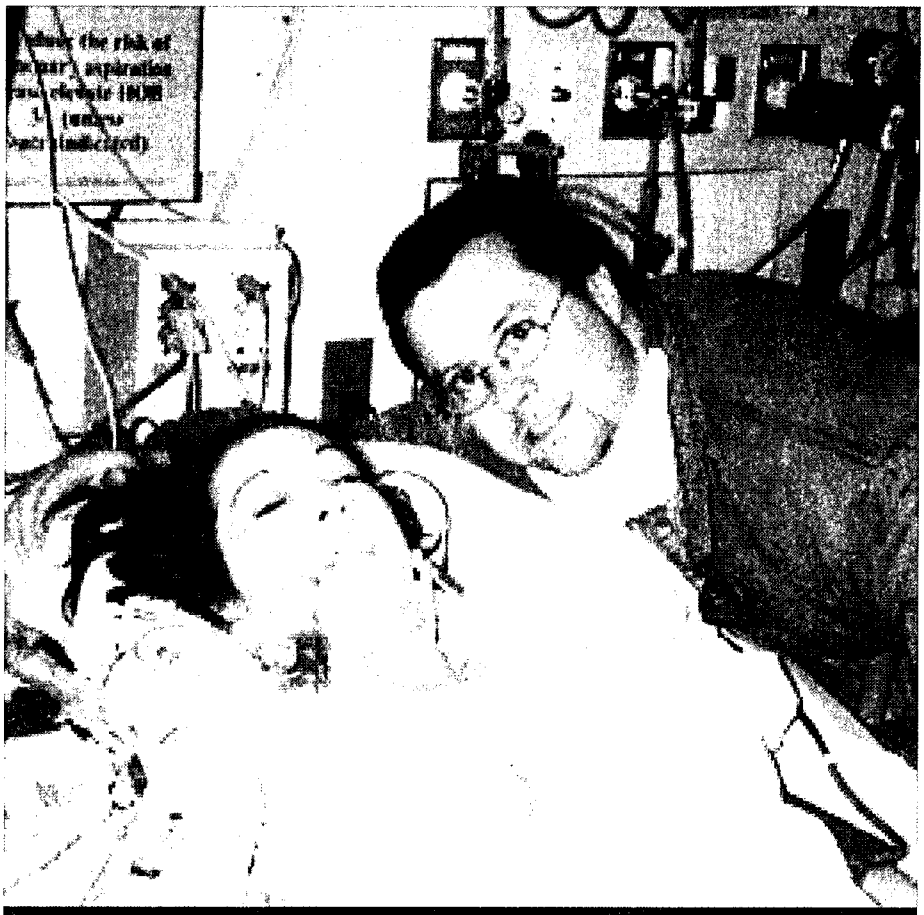
Making the decision

The need for a lung transplant indicates that an individual with CF has reached the late stages of CF, where conventional treatments no longer improve quality of life. Deciding to have a transplant is a significant decision that requires serious consideration because:

- Transplantation is a major surgical procedure that includes a certain level of risk.
- Transplantation requires a life-long commitment to maintaining a rigid post-transplant regimen.

To help make a decision about transplantation, individuals with CF may want to consult with family members, and their CF clinic and transplant teams. A CF clinic and/or transplant team can put potential candidates in contact with a "support network" comprised of other individuals and/or families who have undergone lung transplantation. However, the final decision on whether to proceed with a transplant must be made by each candidate.

Transplant candidates are required to designate a support person (sometimes called a "sponsor") to accompany them to appointments, and to provide support throughout the transplant process.



Margaret Benson

Double-lung transplant recipient: 1999

Post-transplant: spent 18 days in the intensive care unit

2003 and 2005: won gold, silver and bronze medals at two World Transplant Games, in Canada and France

"Deciding to get a lung transplant was difficult for me. I knew that someone would have to die for me to live, and I struggled with that injustice. However, after much consultation and guidance, I was able to make an informed decision: transplantation was a chance I had to take.

If you had told me, when I was at the end-stages of lung disease or when I had a full-body seizure caused by a stroke post-transplant, that I would be winning medals at the World Transplant Games in running and rowing events, I would have thought you were crazy! There were many bumps in my road to recovery. My struggle was not nearly the fight that some have had, but it was up there with the worst of them.

However, these days I often have to think really hard about what it was like to have the lungs of someone with cystic fibrosis. I remain forever grateful to my donor and my donor family who, in a time of tragedy, were able to think of others in need."

Referral and evaluation

Candidates need to go through a pre-lung transplant assessment to determine suitability for a transplant, and to help them to make a decision. Generally, the CF clinic respirologist refers candidates to the lung transplant program. A referral includes transmission of a candidate's complete clinical notes and most recent test results. A transplant centre respirologist reviews the information received and books the candidate for a clinic appointment. During the first appointment, information is given to candidates and their support persons. If the timing is adequate, and an individual is deemed a good candidate, a decision is made by the transplant team on whether to proceed with the complete pre-lung transplant assessment. A referral to proceed with the assessment doesn't mean a candidate has been placed on the transplant waiting list.

Pre-lung transplant assessment

A pre-lung transplant assessment, performed by a transplant team and/or a CF clinic team, includes a number of tests that evaluate lung, heart, kidney and liver functions, as well as an assessment of nutritional status. On average the assessment process lasts one week. Individuals with CF are familiar with many of the procedures, including blood tests, chest X-rays and pulmonary function tests. In addition to assessing physical health, psychological assessments are made to determine a candidate's and his/her family's ability to cope with the stresses of a transplant.

In general, the assessment may include:

Pulmonary Function Test (PFT): measures how the lungs manage airflow.

Chest X-ray: determines the extent of damage to the tissues and heart size.

CT Scan of Chest: looks at the heart and lungs in more detail than a chest X-ray.

Arterial Blood Gas: shows how the lungs are using oxygen.

Ventilation-Perfusion (VQ) scan: shows the amount of blood flow to each lung.

X-ray or CT scan of sinuses: looks for presence of chronic sinus infections and determines the extent of damage to the tissues.

Abdominal ultrasound: evaluates digestive organs, including liver and kidney.

MUGA Scan: evaluates the function of the right and left side of the heart.

2D Echo: evaluates the effectiveness of the heart valves and the pumping action of the heart chambers.

Electrocardiogram (ECG): shows the patterns of electrical waves in the heart.

Thallium Persantine Scan: assesses blood circulation in the heart muscle.

Bone Density Scan: evaluates the strength of the bones.

Sputum analysis: evaluates the types of organisms found within a candidate's airway, and indicates what antibiotics could be used in case of infections.

Six-minute walk test: tests the level of fitness as a candidate walks as fast and as far as he/she can in six minutes, to determine the amount of oxygen required at rest and during exercise.

During a pre-transplant assessment, candidates will also meet with other members of the transplant team. Individual health care issues will determine whether candidates need to see any other specialists.

Assessment meeting

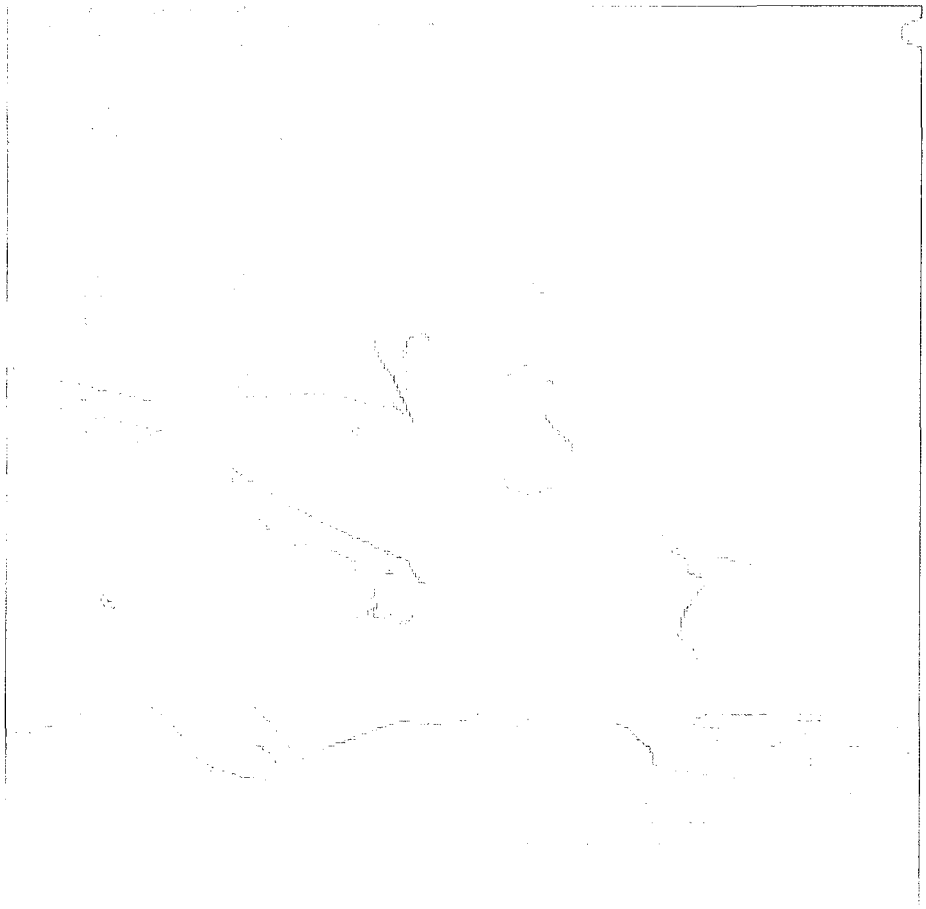
Approximately 2-4 weeks after a pre-transplant assessment is completed, the transplant team (respirologist, thoracic surgeon, transplant coordinator, social worker, dietician, physiotherapist and nurses) meets to discuss the candidate's case, to decide if he or she fits the criteria for transplantation and if the time is right to place this candidate on the transplant waiting list. The candidate must then decide if he or she would like to go ahead with the surgery and be placed on a transplant waiting list.

Listing and waiting for the surgery

If a candidate decides to be put on a waiting list, other requirements may have to be completed. These requirements may include issues related to designating a support person, or securing funding for medication and/or oxygen. Candidates who live far away from their transplant centre may need to relocate closer to the hospital so that their health can be monitored while they wait, and to ensure they can participate in an exercise program. Candidates will then meet with a transplant surgeon to sign the consent forms for the operation and to be officially listed.

Many candidates consider the waiting period the most difficult part of the transplantation process. Prior to being called for a transplant, candidates may experience a variety of emotions, including fear, anxiety and uncertainty. Throughout the process, members of the CF clinic team and the transplant team are available to assist candidates in coping with their concerns.

There is no way to predict when a suitable donor will become available. Waiting times can range from a few weeks, to many months or years. In part, waiting is influenced by a candidate's blood type and by the number of transplants done at a transplant centre. In Canada, the average wait for lung transplant surgery is approximately six to 18 months.



Bob Nesbitt

Double-lung transplant recipient: 1990

Post-transplant: given 30 percent chance of surviving one year

2004: after a lifetime spent beating the odds, celebrated his 50th birthday with a trip to Las Vegas

"I have been granted the most precious gift: a second chance at life. I can breathe without having to think about it. It's a true miracle. No cough, no shortness of breath. Outstanding! It is truly euphoric.

Four hours post-transplant I was writing coherent messages to my wife: 'can't wait to go dancing'."

Preparing for a transplant

To ensure that all candidates are as healthy and strong as possible before their transplantation, they are required to participate in a physical rehabilitation program and to eat a healthy diet. Adequate body weight and good physical health are important, as both can help candidates during surgery, and can also help reduce the risk of complications post-transplant. Pre-transplant exercise can be very important in helping candidates to regain strength after their transplant. Transplant candidates are advised to exercise as much as possible to maintain, or improve, current abilities. A transplant team or CF clinic physiotherapist will design a fitness program that meets with each candidate's needs and abilities. Many transplant centres also offer pre-transplant education programs and support groups. A dietician at the transplant centre or at the CF clinic will devise a diet to help maintain or increase a candidate's weight prior to the surgery.

Donor organs and assignment of organs to candidates

Organs accepted for donation must meet certain criteria to ensure they are in good condition. Meanwhile, a variety of factors are used to determine how organs are assigned; these include, but are not limited to: blood type; size of the available organ; the length of time on a waiting list; and, at some transplant centres, the current degree of urgency. In most cases, lungs become available as the result of a death of an organ donor. Race and gender of a donor or a transplant candidate have no bearing on the match.

Proximity to transplant centre

Because organs cannot survive outside the body for an extended period of time, candidates on waiting lists are sometimes required to find accommodation within a two-and-a-half hour drive/flight (under average conditions) of their transplant centre. As it is impossible to know when the call for surgery will occur, candidates must always be reachable by pager. Transplant centres provide candidates with pagers to ensure they can always be contacted when lungs become available. Please note that some transplant centres are serviced by an air ambulance system that allows candidates to live at home until their transplant surgery.

The call

When suitable lungs become available, candidates will receive a call or a page. As the call may come at any time day or night, candidates are advised to have a readiness plan.

A "dry run" or "false alarm"

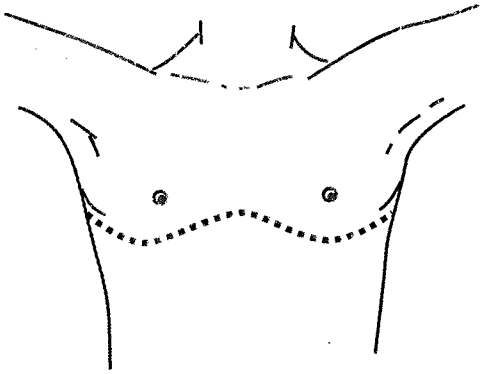
Timing and sequence of events can be critical factors for a successful transplantation. Once lungs have been removed from a donor, transplantation must occur as quickly as possible. Because the condition of donor lungs is not known until the lungs are retrieved, it is possible for candidates to be notified that lungs are available, but to arrive at the hospital to learn that the lungs are not suitable for transplantation. This is known as a "dry run" or "false alarm." Interestingly, many candidates report that having one "false alarm" helped them to prepare for the real call.

Availability of organs

The primary obstacle in obtaining a lung transplant is the availability of suitable donor organs. Organ donation rates in Canada are slowly increasing, but the number of patients listed for transplantation continues to grow at a faster rate.

The surgery

Individuals with cystic fibrosis undergoing lung transplant surgery always receive double-lung transplants to reduce the risk of infection. Lung-transplantation surgery may take anywhere from five to 10 hours. Double-lung transplant surgery is performed through a "transverse sternectomy incision" (across the chest as seen below), and the lungs are transplanted one at a time. Following the operation, lung transplant recipients are placed on a ventilator to assist with breathing, and are moved to the Intensive Care Unit (ICU).



transverse sternectomy incision

Valérie Mouton

Double-lung transplant recipient: 2004

Post-transplant: was left completely deaf, and has developed necrosis of the knees, both as a result of life-saving medicines used to keep her alive while she waited for lungs to become available

2006: returned to university to continue her studies

"I was on the transplant waiting list for 14 months, and I spent 10 of those months permanently hospitalized. At first I was full of hope and enthusiasm, but as my physical strength diminished, my morale began to fade. As I saw my chances for survival becoming slimmer and slimmer, I tried my hardest to have faith. I will never forget the moment when the doctor finally said to me, 'Valérie, your wait is over.'

I only had a few days left to live. If I hadn't had my transplant, I would be dead.

I had quite a few problems after my transplant. The worst part was being deaf. But recently I received a cochlear implant and I can hear again."

Post-transplant

In the ICU, recipients will remain on a ventilator until their new lungs are functioning well, which can take anywhere from one day to many weeks. Visiting the ICU is usually restricted to immediate family members. Once the ventilator is removed, recipients will experience breathing with new lungs. For some individuals with CF, this may be the first big breaths they have taken in many years. Generally after a double-lung transplant, recipients achieve close to normal lung function, but it can take several months for full lung capacity to return.

Transplant recipients must take immunosuppressive drugs, also called anti-rejection drugs, because the body's immune system is programmed to destroy anything foreign and it recognizes the new lungs as foreign tissue. Immuno-suppressive drugs reduce the immune system's ability to attack and reject the new organs. These drugs must be taken every day for the rest of a recipient's life, and will cause a life-long reduction in the ability to fight infection.

There are two types of organ rejection that can occur post-transplant. Acute rejection most often occurs within the first 12 months after transplantation. Anti-rejection drugs help to reduce the chances of acute rejection. However, even when anti-rejection drugs are taken faithfully, recipients can still experience rejection. The transplant team will teach recipients how to recognize and monitor for signs of rejection. It is important to treat organ rejection at the first sign of a problem.

The second type of rejection is called chronic rejection, or *bronchiolitis obliterans (BO)*. Chronic rejection occurs in more than 50 percent of recipients surviving lung transplantation for more than five years, and is the main cause of death in long-term recipients surviving lung transplantation. Chronic rejection may occur gradually, over a long period of time. Chronic rejection is the progressive loss of lung function as a result of inflammation and irreversible scarring of

the smaller airways. Treatment of chronic rejection is very difficult. The goal of treatment is to avoid further decreases in lung function. Treatment may include altering the anti-rejection drugs, and, in very selected cases, re-transplantation.

In the weeks following transplantation, recipients can expect the following:

Monitoring for rejection of organs: Recipients must have regular pulmonary function tests, chest X-rays, and bronchoscopies.

Adjusting to the immunosuppressive drugs: As each transplant recipient is different, different combinations of drugs are required.

Education: It may take time to learn new, post-transplant routines and to learn about new medications and their possible side effects. Some recipients, particularly those who are pancreatic insufficient, develop diabetes after lung transplantation. A nurse and a dietician, knowledgeable about diabetes management, will help recipients who become pancreatic insufficient learn to monitor and control their blood sugar.

Rehabilitation: Transplantation is stressful on the body. For several weeks following discharge from hospital, transplant recipients will usually undergo an exercise rehabilitation program as their bodies gradually regain strength. Soon after recovery from the procedure, recipients can feel a difference in breathing and exercise abilities. Overall, 90 percent of transplant recipients report satisfaction with their decision to undergo the surgery. Survival rates one year after lung transplantation are approximately 95 percent (for persons without *Burkholderia cepacia* complex infections) and 68 percent (for persons with *B. cepacia* complex). Survival rates five years post-transplant are 72 percent (for persons without *B. cepacia* complex) and 39 percent (for persons with *B. cepacia* complex)[1].

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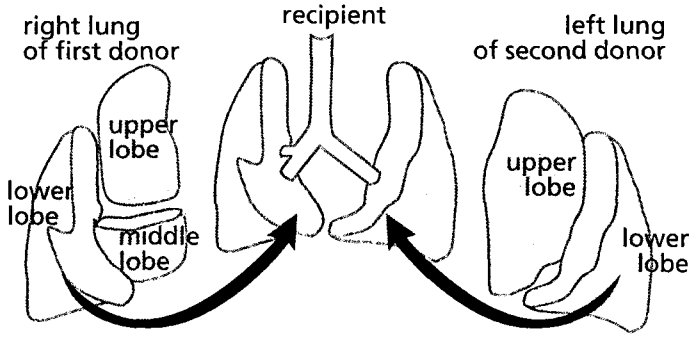
[1] Statistic from The Toronto General Hospital, 1983-2003.

After being released from hospital, follow-up by the recipient's transplant centre and/or CF clinic is fairly rigorous. Initially, recipients attend weekly appointments. These appointments eventually taper off to monthly, and then yearly, as long as a recipient's health is stable.

Living Donor Lung/Lobar Transplantation

In certain circumstances a "living donor lung/lobar transplant," whereby two living donors each provide a lobe from one of their lungs, may be examined. Living donation is considered most often for children and small adults. Potential donors must undergo extensive physical and psychological evaluations; they must have a compatible blood type with a candidate; and donors must be the same height as, or taller than, a transplant candidate so the portion of the lung that is donated is of adequate size.

Currently, living donation is offered only in some transplant centres (Edmonton, Winnipeg, Montreal, and Toronto). Living donors must be in excellent health and must be either a family member, or friend of the recipient. Donors will lose 20 percent of their total lung volume. Typically this loss of lung volume will not affect their health.



The Canadian Cystic Fibrosis Foundation and Lung Transplant Programs

The Canadian Cystic Fibrosis Foundation (CCFF) supports organ and tissue donor awareness and supports every Canadian's decision to sign an organ donor card. In Canada, five institutions host lung transplant programs for individuals with CF. These centres receive Transplant Centre Incentive grants from the CCFF. They include:

- Vancouver General Hospital, Vancouver, BC
- University of Alberta Hospitals, Edmonton, AB
- Health Sciences Centre, Winnipeg, MB
- Toronto General Hospital, Toronto, ON
- Notre-Dame Hospital, Montreal, QC

For help in seeking financial assistance while undergoing transplantation, please contact the transplant team or the Foundation. Information can be found in the Foundation's *Guide to Services, Programs and Resources for the Cystic Fibrosis Community* available through the Foundation or online at www.cysticfibrosis.ca.

Acknowledgements

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For more information about cystic fibrosis or the Foundation, please contact a local CCFF chapter or the Foundation's office:

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2221 Yonge Street, Suite 601
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Telephone: (416) 485-9149
1-800-378-2233
Fax: (416) 485-0960
Web site: www.cysticfibrosis.ca
E-mail: info@cysticfibrosis.ca

CCFF Chapters

There are more than 50 CCFF chapters across Canada. For more information, please contact the Foundation's office or visit the CCFF Web site.

CF Clinics

There are 38 CF clinics across Canada. For a current list, please contact the Foundation's office or visit the CCFF Web site.

In Quebec, you may contact the CCFF's provincial association:

Quebec Cystic Fibrosis Association

Canadian Cystic Fibrosis Foundation

425 Viger Street West, Suite 510
Montreal, Quebec H2Z 1X2

Telephone: (514) 877-6161
1-800-363-7711
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Web site: www.aqfk.qc.ca
E-mail: info@aqfk.qc.ca
Quebec City office: 1-877-653-2086

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Appendix O

Randomized Trial of a Decision Aid for Cystic Fibrosis Patients Considering Lung Transplantation

Katherine L. Vandemheen; Annette O'Connor; Scott C. Bell; Andreas Freitag; Peter Bye; Alphonse Jeanneret; Yves Berthiaume; Neil Brown; Pearce Wilcox; Gerard Ryan; Nancy Brager; Harvey Rabin; Nancy Morrison; Peter Gibson; Mary Jackson; Nigel Paterson; Peter Middleton; Shawn D. Aaron

On-line Data Supplement

Methods

CF Decision Aid

The Ottawa Decision Support Framework was used to guide the development of the CF decision aid (1). The decision aid is a web-based and paper-based booklet that is interactive, self-administered and self-paced. A web-based version of the decision aid is available online at http://decisionaid.ohri.ca/decaids.html.

The decision aid asks the patient to work through five steps when considering the options of "Not to be referred for lung transplantation" or "To be referred for lung transplantation". The pros and cons of each option are presented using illustrative icons for the survival statistics and for the potential long term complications associated with each option.

A separate decision aid was developed for patients infected with *Burkholderia cepacia*. Patients infected with *Burkholderia cepacia* have poorer outcomes post lung transplant (2,3), and these modified survival statistics were presented in the decision aid for *Burkholderia cepacia*-infected patients.

Outcome Measures

The primary end points measured were participants' knowledge, realistic expectations, and decisional conflict evaluated prior to randomization and three weeks after randomization. Knowledge was assessed by a 4-item multiple choice questionnaire designed to determine the patients' understanding of the benefits and risks of referral and lung transplantation or foregoing referral. The response categories were coded as correct or incorrect. The items were summed

and converted to a score ranging from 0 to 100%. Realistic Expectation was assessed with two questions which evaluated the patients' perceptions of the level of risk for surgery and the probabilities of survival with and without surgery, in 100 people like themselves.

There are no validated instruments for the assessments of knowledge and realistic expectation scores for patients considering lung transplant, since the content of the questions is necessarily specific to the topic being studied. However, published tools and standards for assessment of these variables are included as part of The Ottawa Decision Support Framework. The Ottawa Decision Support Framework (ODSF) is an evidence-based, practical, mid-range theory for guiding patients making health or social decisions. It uses a three-step process to:

a. assess client and practitioner determinants of decisions to identify decision support needs;
b. provide decision support tailored to client needs; and
c. evaluate the decision making process and outcomes.

The ODSF has been used to guide the development and evaluation of more than 30 patient decision aids, practitioner decision support resources, and tools to evaluate the quality and outcomes of providing decision support (4-6). The Ottawa Decision Support Framework provides methodology and user manuals to enable investigators to assess patient knowledge and decisional conflict using validated methodology. The tools used to operationalize these variables in the Ottawa Decision Support Framework are available in print, and on-line.

We used the suggested published structure to design our content specific questions. We used the User Manual- Knowledge. © 1995 {updated 2002}. Available at http://c-ohri-dev.ohri.ca/eval.html to design, score and interpret our knowledge outcome questions.

Four knowledge questions were asked as part of the study. Items were given a score value of 1 if the respondent chose the correct answer and 0 if the response was incorrect or the respondent answered 'unsure'.

An example of a knowledge question that we asked in our study was:

Which option has the greatest chance of relieving advanced CF lung symptoms (such as shortness of breath, cough, low energy and poor exercise ability)?

a) Lung transplant
b) Not having a lung transplant
c) Both are about equal
d) I am unsure

The correct answer was 'a'. If the respondent chose 'a' they scored one point, if they chose any other response they scored zero on this item. The four knowledge items were scored to give a total mean score and percentage of correct scores were also calculated for each respondent.

Note that content validity of the questions was established by having our panel of CF and lung transplantation experts review the questions beforehand to make sure they tapped understanding of the key concepts contained in the decision aid. The psychometric properties of the Knowledge questionnaire used in other clinical settings have been published (see website http://c-ohri-dev.ohri.ca/eval.html#Knowledge). The questionnaires have shown internal consistency coefficients (Kronbach's alpha) of 0.82 and 0.83.

For evaluating realistic expectations we used O'Connor AM, User Manual – Realistic Expectations. © 1995 {updated 2002}. Available at http://c-ohri-dev.ohri.ca/eval.html. We used the user manual to design, score and interpret our realistic expectations questions.

An example of a realistic expectation question that we asked in our study was:

If 100 people with cystic fibrosis decide not to be referred for lung transplant, about how many will be alive at 2 to 3 years?

a) Between 1 and 10 people will be alive at 2-3 years
b) Between 11 and 40 people will be alive at 2-3 years
c) Between 41 and 60 people will be alive at 2-3 years
d) Between 61 and 100 people will be alive at 2-3 years
e) I am unsure

For each item, a response is classified as realistic or accurate, if it falls in the range of known probabilities that correspond to a person's health profile. The known range of probabilities is determined from published evidence. Respondents who chose the correct answer of 'c' scored one point, if they chose any other response they scored zero on this item. The two realistic expectation items were scored to give a total mean score, and were also calculated as a percentage.

Note that for these questions the content validity is based on the scientific evidence. Previous evaluations of realistic expectation questionnaires have shown test-retest coefficients of > 0.80 and Kronbach Alpha coefficients of > 0.70.

Decisional Conflict was evaluated using the Decisional Conflict Scale (7), a 16 item scale with 5 subscales: certainty, informed, values clarity, support and effective decision. All questions use a five item response scale. Scores vary from 0 (low decisional conflict) to 100 (high decisional conflict). The instrument is meaningful to healthcare providers with a specified minimal clinically important difference defined; patients scoring 37.5 or more are likely to delay decisions, whereas those with a score of 25 or less tend to make decisions. The decisional conflict scale has been validated and has been shown to be reliable and sensitive to change (1,8,9). It discriminates between different decision support interventions (10,11) and is acceptable to patients.

Secondary outcomes included patient preparation for decision making, choice, and durability of the decision. Preparation for decision making was

assessed using a validated 10-item questionnaire (12) with all questions using a five item response scale. In addition, patients were asked to indicate whether a decision regarding the choice of referral for lung transplantation had been made at baseline and three weeks after randomization. This scale was classified into three categories "not to be referred for lung transplantation"; "unsure"; or "to be referred for lung transplantation'. Durability of the decision was assessed twelve months later by telephone. Participants were asked if they pursued their choice with respect to referral for lung transplantation or if their disease was not yet at the stage of requiring a transplant referral, they were asked if their current choice was the same as it was twelve months ago.

Value congruence was assessed by having patients rate their personal values concerning the importance of the benefits and risks of opting for lung transplantation or foregoing lung transplant referral. We then assessed congruence of these values with the choice made. Results are shown in Table E1.

Acceptability of the decision aid and the materials provided was assessed by asking the patient to evaluate the amount of information they were given, the clarity of the information, the helpfulness of the information and the balance of the options presented using structured response categories (13).

Table Legends:

Table E1: Values Congruence

Table E1: Values Congruence

	Before Randomization		After Randomization	
	OR (CI)	P-value	OR (CI)	P-value
Live as Long as Possible	1.82 (1.04-3.18)	0.04	1.8 (1.02-3.17)	0.04
Avoid Complications	0.59 (0.31-1.13)	0.11	0.66 (0.33-1.29)	0.22
Improve Breathing	1.74 (1.01-3.02	0.047	1.12 (0.67-1.87	0.65
Hassle, Worry, Stress of Transplantation	0.85 (0.7-1.03)	0.09	0.77 (0.6-1.00)	0.047

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