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Caregiver Perspectives on Discussions About the Use of Intensive Treatments in Cystic Fibrosis

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Abstract

Context—Intensive treatments intended to sustain life are often used for patients with advanced cystic fibrosis (CF). There are no guidelines for selecting patients whose survival and quality of life may be enhanced by such treatments or for communication with patients and caregivers about possible treatment outcomes.

Objectives—We aimed to describe caregivers' perspectives on decision making for the use of intensive treatments for patients with advanced CF lung disease.

Methods—We conducted semi-structured interviews with 36 caregivers of 36 patients who died of CF about treatment preference discussions and solicited recommendations for improving discussions.

Results—Twenty (56%) patients received intensive treatments during the last week of life. Twenty-two (61%) caregivers reported ever having discussed intensive treatment preferences with a physician, and 17 (77%) of these discussions were initiated during an acute illness. Only 14 (39%) of all patients participated. Caregivers expressed less certainty about consistency of treatments with patient preferences when patients did not participate. Twenty-nine (81%) caregivers endorsed first discussing treatment preferences during a period of medical stability.

Conclusions—Discussions about preferences for the use of intensive treatments for patients with CF often take place during episodes of acute illness and may be delayed until patients themselves are too ill to participate. Bereaved caregivers suggest first addressing intensive treatment preferences during a stable period so that patient preferences are understood and unwanted treatments are minimized.

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Disclosures

The authors declare no conflicts of interest.

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Keywords

Cystic fibrosis; patient preferences; communication; advance care planning

Introduction

The natural history of cystic fibrosis (CF) is a progressive decline in lung function over time, with intermittent acute exacerbations and, for some, periods of accelerated decline.¹ Although survival in CF is increasing over time,² patients are cognizant of a shortened lifespan for many years preceding death. Intensive treatments, such as noninvasive ventilation or mechanical ventilation, which are intended to sustain life when used during acute respiratory decompensation, are commonly used in patients with advanced CF lung disease.³⁻⁵ Although these treatments were historically thought to have significant morbidity and limited benefit,⁶⁻⁸ more recent literature supports consideration of these treatments on a patient-to-patient basis, with particular attention to their use as a bridge to lung transplantation.^{5,8-11} Without a firm evidence base or guidelines for selecting patients whose survival and quality of life may be enhanced by such treatments, appropriately informing patients of the risks and benefits of treatment options may be challenging. Additionally, the availability of lung transplantation for advanced CF lung disease may complicate patient-physician communication about treatment preferences.¹²

Investigations suggest that CF clinicians lack a framework for initiating effective communication with patients and their caregivers regarding preferences for the use of intensive treatments.^{13,14} Advance care planning to foster alignment of medical care with individual patients' goals and preferences may not be routinely included in the care of patients with advanced CF¹³ despite recommendations for its early incorporation into routine CF care.^{14,15} Little is known, though, about actual practice and whether it corresponds to these recommendations.

Our goal in the present study was to better understand caregivers' perspectives on decision making for the use of intensive treatments for patients with advanced CF lung disease. We interviewed caregivers of patients who died of complications of CF to characterize the timing of discussions about the use of intensive treatments and the frequency of patient participation. We assessed the outcomes of these discussions by inquiring whether treatments received were consistent with stated preferences. We also elicited recommendations for improving discussions about intensive treatment preferences.

Methods

Subjects

We identified caregivers of all patients with CF who died of complications of CF or from bronchiolitis obliterans syndrome after lung transplantation between 1996 and 2006. We included only caregivers of patients who received their primary CF and/or lung transplant-related care at our institution. We mailed letters to 54 caregivers with current contact information requesting participation in a study about treatment preferences and end-of-life care and asked them to return postage-paid letters if they wished not to be contacted by telephone. We did not contact caregivers or conduct interviews within 60 days of major holidays, patients' birthdays, or anniversaries of patients' deaths. Of 54 eligible caregivers, five could not be reached by telephone to follow up the request for participation. Forty-nine were contacted, and 39 (80%) agreed to participate. Of the 10 who declined, seven provided no explanation, two cited sadness related to the patient's death, and one felt uncomfortable participating because of the declining health of another family member with CF. Thirty-six

caregivers, representing 36 caregiver-patient pairs, answered questions about treatment preference discussions sufficiently for analysis.

Interviews

Participants completed a semi-structured interview about the experiences of corresponding patients who died of complications of CF. We adapted questions about end-of-life care from existing bereavement questionnaires¹⁶⁻¹⁸ and developed a series of closed-ended questions addressing whether and when discussions about intensive treatment preferences occurred, whether or not patients participated in these discussions, and whether patient preferences were upheld. Participants were invited to expand on responses to closed-ended questions. This qualitative approach was chosen because of the paucity of data about treatment decision making in CF, a small potential subject pool, and our goal of exploring concepts that might inform the development of future prospective studies about treatment decision making and advance care planning in CF.

The interview script was reviewed by all investigators, approved by our institutional review board, and then pilot tested with three care-givers (two parents and one sibling) of patients who died of CF. One investigator (E. D.) conducted all interviews by telephone after obtaining verbal informed consent from participants. Audiotaped interviews were professionally transcribed. Interviews were designed to last 60 minutes; average length was 54 minutes (range 30–90 minutes). We assessed transcripts for consistency of information reported by care-givers with patient information contained in our CF database and medical records.

Data Analysis

Summary statistics were used to analyze results to closed-ended questions about timing of treatment preference discussions, patient participation in discussions, and consistency of treatments with patient preferences. We categorized responses about timing of first discussions about intensive treatment preferences based on whether these discussions took place during a stable period, including routine clinic visits and during outpatient discussions about referral for lung transplantation, or during acute illness, such as during hospitalization for a severe respiratory exacerbation or when death was felt to be imminent. Differences in categorical variables and trends were compared using χ^2 for larger samples and Fisher's exact test for smaller samples. Missing variables were excluded from the analysis. Expanded responses to closed-ended questions were reviewed for suggestions about improving treatment preference discussions, and representative quotes were selected for inclusion in this presentation of the data.

Results

Caregiver and Patient Characteristics

Caregiver and patient characteristics are summarized in Table 1. Most caregivers interviewed identified themselves as the primary caregiver to their corresponding patients, and those who did not, stated that the patient was independent with regard to disease management. Patients, overall, were seriously ill; 81% used oxygen chronically, many had limited functional capacity and required frequent hospitalizations for respiratory illnesses, and most died of respiratory failure. Most met medical criteria for referral for lung transplantation,^{19,20} and 70% were either awaiting or had undergone transplantation. All lung transplant recipients had developed severe bronchiolitis obliterans syndrome, with lung function in the range where transplanting CF lungs might be considered. A small minority (8%) was offered a transplant and was declined. Eighty-one percent of patients died in the

hospital, and 47% of deaths occurred in intensive care units. Patient deaths occurred an average of 6.7 years (range 1.8–10.9 years) before the interview.

Given the sensitive nature of the topic and the decision to conduct interviews by telephone, we asked the first 12 caregivers whether or not they considered the interview to be an emotional burden and whether they would recommend participation to other bereaved caregivers. Eleven of 12 felt the interview was not an emotional burden, with one stating that it was somewhat of a burden; all 12 said that they would recommend participation in the study to other caregivers.

Participants in Discussions About Intensive Treatment Preferences

Features of discussions about intensive treatment preferences between patients, physicians, and caregivers are summarized in Table 2. Twenty-two (61%) caregivers reported that a discussion with a physician about intensive treatment preferences ever occurred; 14 (39%) of these discussions included the patient, with the remaining taking place between care-giver and physician. Only half of the caregivers reported that the patient had ever discussed intensive treatment preferences directly with them before a discussion taking place with a physician. Patients who had never discussed treatment preferences with a caregiver were less likely to participate directly in discussions with physicians (17%) than those who had (83%; $P < 0.01$); rather, these conversations were limited to physicians and caregivers. Inquiry about other advance care planning revealed that only 37% of adult patients had designated a surrogate decision maker, and one-third had advance directives.

Timing of Discussions About Intensive Treatment Preferences

A large proportion (77%) of discussions about intensive treatment preferences were initiated during acute illness, either while hospitalized for a severe respiratory exacerbation or when death was felt to be imminent. In contrast, 64% of caregivers recommended starting these discussions during stable periods, and an additional 17% felt this was ideal, but that patient and family factors may influence actual timing of discussions. Trends toward discussions occurring during acute illness rather than during stable periods were identified for children, for patients who did not discuss intensive treatment preferences with a care-giver, for lung transplant candidates, and for patients who did not meet lung function criteria for transplant referral. Most caregivers (91%) felt that discussions about intensive treatment preferences should be initiated by physicians.

Consistency of Treatments with Patient Preferences

A minority of patients themselves participated in discussions with physicians about intensive treatment preferences, most often, because, at the time of the discussion, the patient had severe illness with altered level of consciousness or sedation for mechanical ventilation. Thirteen of the 14 (93%) caregivers of patients who participated in treatment preference discussions felt that the patient understood the reason for and content of the discussion very well and that 11 (79%) definitely did not receive unwanted treatments. For the eight patients who did not participate in the discussion, only two (25%) caregivers felt certain that no unwanted treatments were administered ($P = 0.14$).

Overall, 20 (55%) caregivers felt certain that no unwanted treatments were administered, 14 (39%) were uncertain, and two (6%) reported knowing that the patient received unwanted treatments. Fewer caregivers felt confident that no unwanted treatments were administered when discussions occurred during acute illness vs. during stable periods (20% vs. 47%, $P = 0.36$) and when no discussion occurred (41% vs. 84%; $P = 0.03$). Of the 10 patients who never discussed treatment preferences with a physician and whose care-givers were uncertain about their receipt of unwanted treatments, six were sedated for mechanical

ventilation, and the remaining four were unable to communicate effectively because of respiratory distress and altered mental status. Both caregivers who knew that their corresponding patients received unwanted treatments reported feeling pressure from physicians to pursue these treatments, with one stating: “[Physician] looked at mechanical ventilation like the magical thing.”

Caregiver Recommendations for Discussions About Intensive Treatment Preferences

Most of the caregivers endorsed initiating discussions about intensive treatment preferences during a period of stability. All who felt that appropriate timing depended on patient characteristics favored discussions during stable periods, but some pointed out that this may not always be possible because of unanticipated life-threatening illness. Others noted that some patients and caregivers are not well-equipped to participate in such discussions because of lack of understanding, fear of death, or denial of the severity of illness or of disease prognosis. One caregiver commented, “You can’t wait until somebody’s on death’s door. And you can’t talk to them today and expect a decision.” Another caregiver who noted the importance of patient preferences being known to all members of the medical team said, “... So if certain decisions have already been discussed ... and it was done deliberately and wasn’t something that had to be quickly started on the spot, ... [patient] could really talk it through and come to that decision.”

Many caregivers offered suggestions for improving communication or timing of treatment preference discussions. Some suggested providing more information to patients and caregivers about indications for and potential outcomes of intensive treatments. Of note, nine caregivers mentioned knowing that outcomes of mechanical ventilation in CF are often poor, but only four recalled hearing this from a physician, whereas the remaining five received this information from other patients and caregivers. Some caregivers remarked that addressing treatment preferences first during acute illness is not ideal; hospitalizations or times of decline in health are appropriate times to readdress preferences. Many mentioned that decisions are often affected by situational factors, with one caregiver of a patient with whom no discussion occurred stating, “[Have these discussions] early instead of getting close to when you might be depressed or desperate just because you’re facing dying.”

Most caregivers felt that patients should participate in discussions so as to enhance their understanding of treatment decisions; delaying discussions until patients are too ill to participate may put caregivers in an uncomfortable position and place patients at risk of receiving unwanted treatments. One caregiver said, “... I wish that I had had time to talk to him, but I didn’t. The doctor said we have to intubate him now or he is going to die. And I hated that because it scared me. I am sure it scared him. And I had to make a decision very quickly but I wanted to talk to his father, so I was running around trying to call him. And the panic that you have because you need to make this decision quickly.” Another said, “... I feel like maybe there should have been a discussion before she got so ill that she couldn’t talk about it, so that we could have spoken with her about it so that the doctors would know. Because I truly don’t think those doctors truly understood how she felt.”

Most caregivers felt that the primary CF physician should initiate discussions about intensive treatment preferences. Three caregivers expressed anger about physicians not initiating discussions, noting that these are difficult issues for patients and caregivers and that by “opening the door,” as one caregiver stated, patients and caregivers can then help guide the direction of discussions. Although many acknowledged the difficult nature of these discussions, they also pointed out that patients and caregivers are aware that CF is life limiting; thus, discussing treatment preferences is necessary and appropriate. One stated, “... inevitably they [patients] are going to realize that this is a life-threatening disease ... unless they’re totally unable to talk about it or don’t comprehend, they are going to wonder, what’s

my life going to be like? And I think picking up on those cues would be important as early as possible.”

Discussion

As intensive treatments intended to sustain life are often used for patients with advanced CF despite a lack of guidelines for selecting patients who may derive benefit or a framework for communication with patients and care-givers about possible treatment outcomes, we sought to understand caregivers’ perspectives on treatment decision making. We found that discussions about preferences for the use of intensive treatments often take place during acute illness and late in the disease course, and that patient participation may be affected by the timing of these discussions. Discussions during acute illnesses rather than during stable periods may be even more common for younger patients, those who await or have undergone lung transplantation, and those who do not communicate about treatment preferences with their caregivers independently of discussions with physicians. The caregivers we interviewed endorsed discussing intensive treatment preferences during periods of medical stability and readdressing preferences over time. The vast majority felt that physicians, not patients or caregivers, should initiate discussions about treatment preferences.

Medical care for patients with advanced CF lung disease often incorporates more intensive treatments and preventive treatments than those used in the traditional model of palliative care.^{14,21} Many factors likely influence the utilization of intensive treatments, including, but not limited to, the availability of lung transplantation, the young age at death of many patients, lifelong experience of frequent medical interventions, and the fact that many patients with CF die in the hospital.^{3,22,23} The anticipated course is a gradual downward trajectory in lung function, but episodes of acute illness and severe exacerbations from which patients do not return to their previous state of health are difficult to predict, and a terminal phase may be difficult to recognize for physicians, patients, and caregivers alike. Although predictive models of survival and lung transplant outcomes in CF exist,^{24,25} their appropriate application to clinical practice is uncertain. All of these factors may make the appropriate timing of discussions about intensive treatment preferences difficult to determine.

Incorporation of advance care planning into routine CF care is recommended,^{14,15} and initiating discussions about intensive treatment preferences and other advance care planning earlier and more systematically could allow patients to make more informed choices and achieve better palliation of symptoms.²⁶ Additionally, the care provided to patients with advanced disease could be more consistent with patient and caregiver goals, with unwanted treatments minimized and desired treatments offered when felt to be appropriate. Although physicians who care for patients with CF may not routinely address the various aspects of advance care planning, our study suggests that patients and caregivers want and expect physicians to initiate discussions about the use of intensive treatments. Perhaps more advance care planning would follow from a different strategy for communicating with patients and caregivers.

For physicians to develop strategies for addressing these issues with patients and caregivers, they must recognize that discussing treatment preferences with patients fosters communication between patients and their caregivers and relieves some of the burden of surrogate decision making often placed on caregivers.²⁷ Physician practices for discussing intensive treatment preferences with CF patients and their caregivers have not been described, and there is no common framework for initiating effective communication with patients and their caregivers regarding treatment preferences.

There are a number of limitations to this study. First, this is a single-center study; hence, it does not address practices at other CF-care centers. We only interviewed caregivers whose corresponding patients died, and the views of caregivers of living patients may be different. We asked caregivers to recall details of discussions that took place many years before the interview. Because CF is a relatively uncommon disease, the only way to collect adequate numbers of participants is to include patients over time. To assess accuracy of responses, we compared recalled events with factual demographic and disease-specific information previously abstracted from medical records and found concordance. Using caregivers as surrogates for patients may misrepresent actual events or the feelings and opinions of patients themselves, but as caregivers are closely involved in the care of many patients with CF even into adulthood,²⁸ including making complex medical decisions, their perspectives are useful in addressing the study questions. Additionally, use of surrogates to study end-of-life experiences²⁹ and to inform advance care planning is the most feasible method to collect data on patients' experiences in the final moments of life.³⁰

Despite these limitations, the findings of our study suggest a need for further exploration in the areas of communicating about treatment preferences and advance care planning in CF. Further studies of physician practices and prospective studies of patients and caregiver experiences may help to provide a framework for such discussions. The impact of lung transplant on treatment decision making also warrants further attention. Improving communication about these important issues will allow for more concordance between patient and care-giver goals and the care provided by physicians.

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Table 1Caregiver and Patient Characteristics ($n = 36$ Caregiver-Patient Pairs)

Caregiver and Patient Characteristics	Number (% of 36 Total Caregivers) Unless Otherwise Indicated
<i>Caregiver characteristics</i>	
Median age in years (range)	57 (32–82)
Relationship with patient	
Parent	31 (86)
Spouse	2 (6)
Sibling	1 (3)
Grandparent	1 (3)
Friend	1 (3)
Female gender	32 (89)
Primary caregiver to patient	30 (83)
<i>Patient characteristics</i>	
Median age at death in years (range)	23.7 (8–47)
Adult (age ≥ 18 years)	24 (67)
Frequency of hospitalizations during last 6 months of life	
Monthly	10 (28)
Every other month	13 (36)
Once or twice	12 (33)
None	1 (3)
Level of function during last 6 months of life	
Very limited	11 (31)
Somewhat limited	17 (47)
Not very limited	5 (14)
Not limited at all	3 (3)
Lung transplant status	
Awaiting transplant	6 (17)
Underwent transplant	19 (53)
Declined transplant	3 (8)
Not referred for transplant ^a	8 (22)
Cause of death	
Respiratory failure	33 (92)
Septicemia	3 (8)
Location of death	
Hospital—medical ward	12 (33)
Hospital—intensive care unit	17 (47)
Home	7 (19)
Intensive treatments used in the last week of life	
Mechanical ventilation	14 (39)
Noninvasive ventilation	6 (17)
Cardiopulmonary resuscitation attempted ^b	4 (11)

^aOf those not referred for transplant, three did not meet medical criteria for referral. Of the five who did, three were not referred because of difficulties with adherence to therapies and two because of medical contraindications to transplant.

^bAll patients also were receiving mechanical ventilation.

Table 2**Patient-Physician Discussions About Intensive Treatment Preferences and Other Advance Care Planning: Caregiver Responses**

Characteristic of Discussion or Issue Discussed	n (%)
Patient discussed intensive treatment preferences with caregiver	18 (50)
Discussion with a physician about intensive treatment preferences occurred	22 (61)
Patient definitely participated in discussion with a physician	14 (39)
Timing of first discussion about intensive treatment preferences	
During a period of stability	5 (23)
During an acute illness	17 (77)
Caregiver's perception of ideal time for first discussion to take place	
During a period of stability	23 (64)
During an acute illness	7 (19)
Appropriate time depends on patient and caregiver characteristics	6 (17)
Caregiver's perception of who should initiate discussions about intensive treatment preferences	
Primary CF physician	33 (91)
Patient or family member	2 (6)
Social worker	1 (3)