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Effects of lung transplantation on inpatient end of life care in cystic fibrosis

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Abstract

Background—The impact of lung transplantation on end of life care in cystic fibrosis (CF) has not been widely investigated.

Methods—Information about end of life care was collected from records of all patients who died in our hospital from complications of CF between 1995 and 2005. Transplant and non-transplant patients were compared.

Results—Of 38 patients who died, 20 (53%) had received or were awaiting lung transplantation ("transplant" group), and 18 (47%) were not referred, declined transplant, or were removed from the waiting list ("non-transplant"). Transplant patients were more likely than non-transplant patients to die in the intensive care unit (17 (85%) versus 9 (50%); P=0.04). Sixteen (80%) transplant patients remained intubated at or shortly before death, versus 7 (39%) non-transplant patients (P=0.02). Do-not-resuscitate orders were written later for transplant patients; 12 (60%) on the day of death versus 5 (28%) in non-transplant patients (P=0.02). Transplant patients were less likely to participate in this decision. Alternatives to hospital death were rarely discussed.

Conclusions—Receiving or awaiting lung transplantation affords more aggressive inpatient end of life care. Despite the chronic nature of CF and knowledge of a shortened lifespan, discussions about terminal care are often delayed until patients themselves are unable to participate.

Cystic fibrosis (CF) is a lifespan-limiting genetic disease with predicted median age of survival of 36.8 years in the United States.1 The natural history of CF is a progressive decline in lung function secondary to chronic infection, with death resulting from respiratory failure in most patients.2 Advancements in CF care, including improved pulmonary and nutritional therapies, have led to improved survival over time. In addition, lung transplant is

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a therapeutic option which may improve survival3 and quality of life3,4,5 for some patients. Since the reality of the disease is that it generally leads to death at an early age, physicians who provide care for CF patients should be prepared to help manage both the medical and psychological aspects of care at the end of life.

It has been suggested that end of life care in CF does not fit the traditional model of palliative care.6 CF patients are more likely to die in the hospital than at home or in hospice care facilities, and tend to receive aggressive medical care.7 Whether death typically occurs in the acute care setting because of patient wishes or because of lack of alternatives has not been described. Lung transplant patients who do not survive may be even more likely to die in the hospital, specifically in the intensive care unit (ICU).8 This may be related to assumptions medical providers make about patient desires for treatments at the end of life. The aims of this study were (1) to assess various aspects of end of life care in patients dying from complications of CF, (2) to determine whether differences exist with regard to end of life care in transplant and non-transplant patients, and (3) to assess whether alternatives to death in the acute care setting are discussed and offered to CF patients.

Methods

We reviewed the medical records of all 119 patients who died from complications of CF between November 1995 and November 2005 while under the care of the University of North Carolina Cystic Fibrosis Center (Figure 1). Seventy-seven of these patients received CF-related care at our center for a minimum of two years preceding death. Little information was available about the 33 patients who died outside the institution. Many of these patients lived a great distance from our center, thus were more likely to have died in hospitals in their communities rather than at our institution. Forty-four of the remaining patients died as inpatients at our institution during this period, and these were the patients in whom there was sufficient information documented about end of life care to complete the chart review.

From this group of 44 patients, 6 were excluded because they either did not survive to hospital discharge following lung transplant (5 patients) or had undergone lung transplant within the preceding six months (1 patient), a factor which would likely influence decision-making about end of life care. Information about the remaining 38 patients was included in the analysis. These 38 patients were divided into two groups based on their lung transplant status. Twenty patients (53%) who had either received or were awaiting lung transplant comprised the “transplant” group. These patients were grouped because of presumably similar exposures to discussions about lung transplantation and to care by our transplant team, which differs from the CF center medical team. The “non-transplant” group included 18 patients (47%) who were not referred for transplant, were considered for transplant but subsequently removed from the transplant list, or declined transplant (Table 1). Patients younger than 18 years were considered “children” for the analysis.

Medical records were reviewed for the following: basic demographic information; disease-specific information such as lung function and respiratory pathogens; lung transplant status; and information concerning end of life care, including setting of death, the use of

mechanical ventilation, documentation of advanced directives, timing of do-not-resuscitate (DNR) orders, and patient involvement in discussions about terminal care.

For the purposes of this study, the term “advance directives” was defined as previously-discussed wishes for life-sustaining treatments, and implies direct communication with the patient regarding such wishes. “Final resuscitation status” was defined as the resuscitation status documented in the medical record which determined whether or not resuscitation was attempted in the event of a cardiopulmonary arrest. “Full code” status was defined as desiring an attempt at resuscitation in the event of a cardiopulmonary arrest, and a “DNR” status was defined as desiring no intubation or resuscitation efforts. Those who remained intubated at the time of death or until shortly before, such that their death occurred due to withdrawal of ventilatory support, were considered “intubated” for the analysis.

For this descriptive study, summary statistics were used to analyze all study variables. Differences in categorical variables and trends were compared using X^2 for larger samples and Fisher's exact test for smaller samples. For categorical variables with more than two categories, P-value for X^2 with more than two degrees of freedom was calculated, and individual pairs were tested in subsets using X^2 or Fisher's exact tests. Missing variables were excluded from the analysis.

The study was approved by the biomedical institutional review board of the University of North Carolina, and was conducted according to its guidelines for protection of patient confidentiality.

Results

Patient Characteristics

For the study population as a whole, the mean age at death was 22.9 years (range 8 to 41 years). Fourteen (37%) patients were children (age < 18 years). The FEV_1 was less than 30% predicted, a factor often used in considering patients for lung transplant,7 in 29 patients (76%) overall. The most common primary respiratory pathogen was *Pseudomonas aeruginosa* (73%), and *Burkholderia cepacia* complex organisms were cultured in 22% of patients. Respiratory failure was the cause of death in 31 (82%) patients overall, with sepsis syndromes responsible for the remainder of deaths.

The patients were divided into “transplant” and “non-transplant” groups as previously described. There were 6 children and 14 adults in the transplant group, and 8 children and 10 adults in the non-transplant group. Comparisons between the two groups are shown in Table 2. The groups did not differ substantially in terms of their basic characteristics except for education, with significantly more transplant patients having enrolled in or graduated from college than the non-transplant group. The mean time between transplant and death was 4.1 years (range 0.6-10.1 years).

Level of Care at the End of Life

Characteristics of end of life care in both transplant and non-transplant patients who died in the hospital are summarized in Table 3. Transplant patients were significantly more likely to

die in the ICU (85% versus 50%; P=0.04) and to be intubated at or shortly before the time of death (80% versus 39%; P=0.02) than non-transplant patients. Non-invasive ventilation was used in 3 non-transplant patients, 2 of whom died on a hospital ward and 1 in the ICU. No transplant patients were using non-invasive ventilation at the time of death. Of those who died in the ICU, all but 1 (94%) of the transplant patients were intubated at the time of death or shortly before, versus only 2 (29%) of the non-transplant patients (P<0.01). Age, gender, marital status or family situation, family history of CF, and a history of prior CF deaths in the family were not independently associated with setting of death or intubation status. A higher level of education was associated with increased likelihood of dying in the ICU and being intubated at or shortly before death, consistent with the fact that there were more patients who had achieved a higher level of education in the transplant group.

Advance Directives and Resuscitation Status

Discussions regarding advance directives were documented in a similar proportion of transplant and non-transplant patients (70% versus 67%; P=0.83). The timing of such discussions was highly variable. Transplant and non-transplant patients differed with regard to desired resuscitation status, with transplant patients more often designating a full code status (60% versus 11%; P<0.01). Transplant patients were less likely than non-transplant patients to be undecided with regard to desire for resuscitation (10% versus 28%; P<0.01).

At the time of death, all but 3 patients (all 3 of whom were awaiting first lung transplant) had a DNR status. There was a trend toward more transplant than non-transplant patients dying via withdrawal of ventilatory support (45% versus 28%; P=0.13) following the decision to establish a resuscitation status of DNR, with the remainder of both groups (with the exception of the 3 transplant patients in whom resuscitation was attempted) dying after a DNR order was written but not requiring withdrawal of invasive support. Transplant patients were more likely to have a DNR order written on the day of death than before the day of death (60% versus 28%; P=0.05). None of the transplant patients had a DNR order written at the time of admission to the hospital.

Comparison of advance directives to final resuscitation status revealed that the 3 patients in whom resuscitation was attempted at the time of death had desired a full code status at the time advance directives were determined. All 5 who had designated a DNR status maintained this status at the time of death. Eleven patients who had designated a full code status were ultimately changed to DNR status. All 19 patients in whom advance directives were not documented or no decision had been made ultimately had a DNR status.

Documentation of patient involvement in discussions about wishes for treatments at the end of life and determining their final resuscitation status was examined. Transplant patients were less likely to be involved in such discussions (15% versus 61%; P<0.01). There was less documentation of such discussions in the transplant group (35% versus 61%; P<0.01), most likely because more of these patients were intubated and sedated, thus unable to participate. When discussions were documented, adults and children were equally likely to be involved, but the actual frequency of documentation of whether or not children themselves were involved in such discussions was quite low (3 children (21%), versus 15 adults (63%); P<0.01).

Alternatives to Inpatient Care

Hospice care was not utilized by any of the patients in the study population as all died in the acute care setting. This option was discussed with 1 adult transplant patient and 1 adult non-transplant patient, both of whom died outside of the ICU. Interest in this option was expressed by the transplant patient, but death occurred before arrangements could be made. The other patient expressed a preference to die in the hospital.

Discussion

It has been reported that patients with CF often receive their end of life care in the hospital, and tend to receive aggressive care.7 This study supports these findings, and provides new insight by demonstrating that differences exist between transplant and non-transplant patients with regard to end of life care. At our center over the period studied, transplant patients were more likely to die in the ICU and to be treated with mechanical ventilation. They were more likely to desire attempts at resuscitation, and were less likely to participate in discussions which precluded this from happening during a severe, terminal illness. They were less likely to be able to communicate effectively with medical providers and with family members and friends around the time of death. As therapeutic care is a component of palliative care in patients with CF,6 and, in some cases, lung transplant remains a viable option until other major complications of CF or previous lung transplant ensue, a more aggressive approach to care may be appropriate. This direction of care, if chosen, should not necessarily limit patient participation in discussions about preferences for treatments at the end of life.

Our study has limitations which must be considered when interpreting the findings. First, our study is a retrospective chart review, and relies on documentation by physicians and other CF care providers. Thus, it only reflects the perspective of certain members of the medical team. In addition, the end of life care of a large number of patients who died under the care of our CF center was not documented at all, as it occurred outside of our institution. We cannot account for the circumstances of death of these patients. Thirdly, we presume that patients self-select for transplant to some degree, such that our comparison groups (transplant versus non-transplant) are not equivalent. Thus, our statistical comparisons are for descriptive purposes only.

Another limitation is that our institution is a lung transplant center which accepts referrals from a large regional territory as well as from across the country, and has in the past offered lung transplantation to some patients infected with *Burkholderia cepacia*, a pathogen that confers increased mortality in CF 10,11 and is associated with poorer lung transplant outcomes.12-14 Thus, this population may not be representative of the overall population of patients with CF dying in the United States. The high rate of infection with *Burkholderia cepacia* in our population and the propensity for infection with this organism to cause more severe illness may have affected the level of care offered to some our patients, whether or not they were candidates for or recipients of lung transplant.

Although the outcomes of patients with CF who undergo invasive therapies for respiratory failure has historically been poor,15 these outcomes may be improving over time, such that

this level of care may be felt to be appropriate under some circumstances.16-18 Patients who have survived previous illnesses involving the use of invasive or non-invasive mechanical ventilation may, in particular, be more willing to undergo such treatments. During the time period reviewed, non-invasive ventilation has been described as a therapy and possible bridging mechanism to lung transplant in patients with CF and acute respiratory failure, 19,20 but this strategy was not utilized in our population as all patients in this category were too ill for this therapy to be effective. In our analysis, no obvious change in level of care over the period of time studied was apparent.

Despite these limitations, our findings have important implications and suggest further areas of study. One such area is when to address preferences for treatments at the end of life. Patients with CF have known, often for a large portion of their lives, that they have a lifespan-limiting disease, but there are many potential barriers to communication about end of life issues. For example, the hope offered by improved therapies and lengthening of the lifespan in patients with CF may affect the ability and desire of both patients and physicians to have such discussions. Aggressive management strategies such as lung transplant may seem inconsistent with discussion of end of life care. Recent data that brings into question whether lung transplant improves survival in certain patients14 may prompt CF physicians to reconsider their approach to discussions about transplant. It is important to discuss transplant outcomes in terms of both survival and quality of life. Preferences for treatments at the end of life are a logical inclusion in such discussions, but as evidenced by our population of dying patients, if discussions about end of life care are not held before the time of transplant referral, a substantial proportion of patients may not benefit from such discussions. Enhancing communication about such issues may be beneficial for patients, families and providers alike, and may facilitate the development of models of end of life care for patients with CF.21

Investigation of alternatives to dying in the hospital is also warranted. Studies of inpatient end of life care for other diseases have revealed discordance among patients, families and physicians with regard to symptom assessment and management22-24 and preferences for life-sustaining treatments.24 In addition, those who experience the death of a hospitalized family member often express dissatisfaction with care at the end of life.23,25,26 Given that many patients with CF die in the hospital, they may be at risk for receiving unwanted or feeling dissatisfied with end of life care. These risks may be even greater for lung transplant patients. Although not studied in CF, it is well-established in many other conditions that patients prefer to die at home or in a hospice facility than in the acute care setting.27-30 Home care for acute CF-related illnesses is commonly practiced,31-36 and hospice services are becoming more widely available for patients with other chronic diseases.37,38 In general, however, the populations served by hospice are older and the diseases leading to their deaths, even when chronic, are not comparable to CF. Despite these differences, it is possible death in the acute care setting is not preferable for all patients with CF. Barriers to home care and referral to hospice programs may exist. It is important for physicians to assess the wishes of their patients at a time when patients are able to consider and investigate their options and to express their wishes.

Conclusion and Authors' Recommendations

In summary, we have found that patients with CF who die in the hospital, particularly those who have undergone or are awaiting lung transplant, receive aggressive care at the end of life and often are unable to participate in discussions and decision-making surrounding end of life care. Given the limited published information in this area, examining these issues at other centers and comparing experiences may provide useful insights. Based on our findings and experience, we recommend the following:

1. Caregivers should document patient and family preferences for end of life care and the use of life-sustaining treatments in a readily accessible section of the medical record.
2. It may be helpful to begin to address preferences for life-sustaining treatments as a part of routine care before patients become terminally ill, as well as at times of deterioration in health and as part of the lung transplant referral process.
3. Care centers should develop strategies to enhance informed decision making and communication. Patient, family and medical provider satisfaction should be evaluated before and after interventions to help assess the value of these strategies.
4. Further research is needed regarding the important factors in decision making about end of life care in CF, both from the perspectives of patients and of the health care team.

Seeking the perspectives of patients and families would help characterize their wishes regarding end of life care and the effects that discussions about end of life care might have on perceived quality of life, compliance with therapies, and decision-making regarding lung transplant. Assessing physician approaches to discussions about end of life care in conjunction with the information offered by patients and families may help determine the optimal timing and appropriate content of such discussions. This information could be used to improve the quality of care we offer to patients with CF at the end of life.

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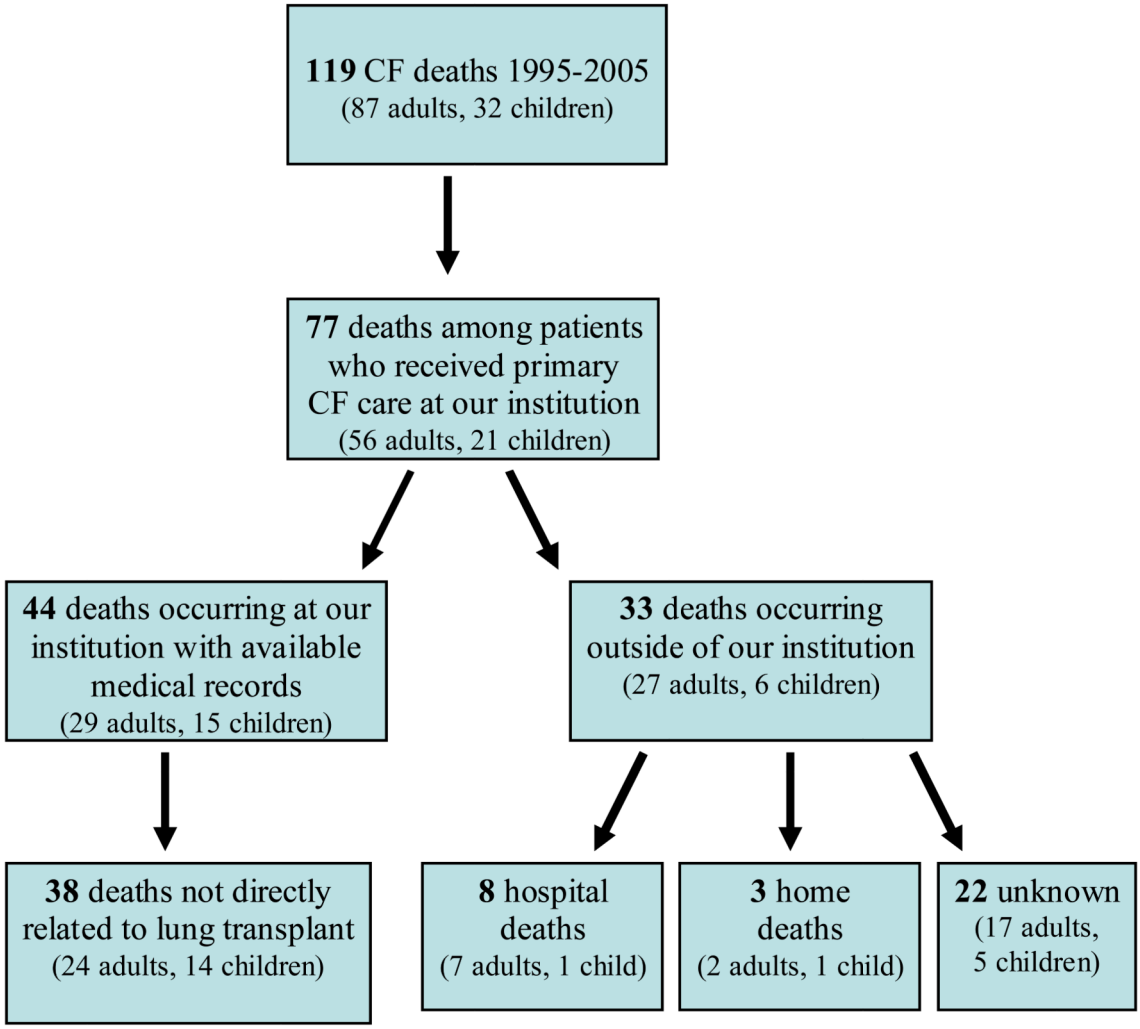

FIGURE 1.
Selection of the study population.

TABLE 1

Classification of Patients According to Lung Transplant Status

Transplant Status	Number of subjects (%)
"Transplant"	20 (53%)
Awaiting first transplant	10
Received one transplant, listed for second	6
Received two transplants	4
"Non-transplant"	18 (47%)
Not referred for transplant[a]	14
Removed from transplant list[b]	1
Declined transplant[c]	3

[a] Seven did not meet criteria for referral, 4 were not referred because of nonadherance to medical therapies, and 3 were not referred for undocumented reasons.

[b] Removed from transplant list because of nonadherance to medical therapies

[c] None had previously undergone a lung transplant.

TABLE 2

Comparison of Characteristics of Transplant and Non-Transplant Patients

Characteristic	"Transplant" n=20	"Non-transplant" n=18	P value[a]
Mean age at death (years)	24.1 +/-8.8	21.6 +/-10.1	0.42
Children - no. (%)	6 (30)	8 (44)	0.36
Gender - Male - no. (%)	7 (35)	8 (44)	0.55
Race - Caucasian – no. (%)	20 (100)	18 (100)	--
Genotype - no. (%)			
Homozygous deltaF508	14 (70)	9 (50)	0.37
Other	6 (30)	9 (50)	
Primary pathogen - no. (%)			
Pseudomonas aeruginosa	16 (79)	12 (67)	0.67
Burkholderia cepacia complex[c]	3 (16)	5 (28)	
Other	1 (5)	1 (5)	
Lung function - no. (%)			
FEV_1 <30% predicted	17 (85)	12 (67)	0.35
Unknown[b]	--	1 (5)	
Cause of death - no. (%)			
Respiratory failure	15 (75)	16 (89)	0.27
Sepsis	5 (25)	2 (11)	
Distance from center - no. (%)			
In-state, less than 100 miles	10 (50)	10 (56)	
In-state, more than 100 miles	7 (35)	7 (39)	0.64
Out of state	3 (15)	1 (5)	
Educational level (adults) - no. (%)			
Less than high school	1 (7)	2 (20)	
High school degree	1 (7)	6 (60)	0.02
College enrolled or graduate	8 (57)	2 (20)	
Unknown[b]	4 (29)	--	
Marital status (adults) - no. (%)			
Married	6 (43)	4 (40)	
Divorced or separated	2 (14)	--	0.41
Single	6 (43)	6 (60)	
Family situation (children) - no. (%)			
Intact family	3 (50)	4 (50)	--
Single parent	3 (50)	4 (50)	
Other CF in family - no. (%)			
Yes, sibling	3 (15)	3 (17)	
Yes, other relative	3 (15)	4 (22)	0.86
Unknown[b]	4 (20)	2 (11)	
Prior CF death in family - no. (%)			

Characteristic	"Transplant" n=20	"Non-transplant" n=18	P value[a]
Yes, sibling	2 (10)	1 (5)	
Yes, other relative	--	4 (22)	0.16
Unknown[b]	5 (25)	3 (17)	

[a] Overall P value, determined by X^2 test

[b] Unknown implies not documented

[c] Prevalence of *B.cepacia* complex was 6.6% at our center in 2005, and prevalence of *B.cepacia* Genomovar III was 3.6%.

TABLE 3

Characteristics of End of Life Care in Transplant versus Non-Transplant Patients Who Died During Hospitalization

Characteristic of End of Life Care	"Transplant" (n=20)	"Non-transplant" (n=18)	P value[a]
Setting of death - no. (%)			
Hospital ward	3 (15%)	9 (50%)	0.04[b]
Intensive care unit	17 (85%)	9 (50%)	
Intubation status - no. (%)			
Not intubated	4 (20%)	11 (61%)	0.02[b]
Intubated	16 (80%)	7 (39%)	
Advance directives - no. (%)			
Unknown or not documented in chart	6 (30%)	6 (33%)	
Full code	12 (60%)	2 (11%)	<0.01[c]
Do not resuscitate	0 (--)	5 (28%)	
Discussed but no decision made	2 (10%)	5 (28%)	
Final resuscitation status - no. (%)			
Resuscitation attempted	3 (5%)	0 (--)	
DNR order written, passed away	8 (40%)	13 (72%)	0.07
DNR order written, intensive support withdrawn	9 (45%)	5 (28%)	
Patient participated in discussion regarding final resuscitation status – no. (%)	13 (65%)	7 (39%)	
Unknown or not documented in chart	3 (15%)	11 (61%)	<0.01[d]
Yes	4 (20%)	0 (--)	
No			
Timing of DNR order - no. (%)			
At/before hospital admission	0 (--)	2 (11%)	
Before day of death	5 (25%)	11 (61%)	0.02
On day of death	12 (60%)	5 (28%)	
Never (full code)	3 (15%)	0 (--)	

[a] Overall P value, determined by X^2 test unless otherwise noted

[b] Fisher's exact test

[c] Transplant and non-transplant patients differed with regard to desiring a "full code" status versus a DNR status (P <0.01) and desiring DNR status versus being undecided (P <0.01), with transplant patients more likely to desire a full code and less likely to be undecided.

[d] Transplant and non-transplants patients differed with regard to involvement in decisions about their ultimate resuscitation status. Transplant patients were less often documented to be involved in the discussion (P <0.01).