Advance Care Planning in Adults with Cystic Fibrosis

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Abstract

Background: Because many patients with cystic fibrosis (CF) continue to survive into adulthood, discussion of end-of-life care decisions between clinicians and patients becomes a crucial part of CF adult care. Advance care planning (ACP) promotes alignment of patient care at the end of life with an individual's goals, however minimal research exists on ACP in CF.

Methods: We surveyed adults enrolled in the Project on Adult Care in Cystic Fibrosis (PAC-CF). We assessed experiences with ACP processes and communication and sought to identify factors associated with completion of an advance directive.

Results: The mean age of respondents ($n = 234$) was 34 years and the mean forced expiratory volume in 1 second (FEV₁) was 64% predicted. Seventy-four percent reported that they had spoken to someone, generally a family member, about the care they would want if they became too ill to make decisions for themselves. However, only 30% reported completing an advance directive. Although 79% reported feeling comfortable talking to their clinician about ACP, only 28% said that their CF clinicians have asked about ACP. Having specific wishes about treatment decisions (odds ratio [OR] 7.8, 95% confidence interval [CI] 1.9–32.1) and reporting that a clinician had discussed ACP (OR 4.4, 95% CI 1.5–12.6) were significantly associated with reporting the completion of an advance directive.

Discussion: Though the majority of adults with CF report thinking about and communicating with family about advance care wishes, only a minority report completing an advance directive. Few adults with CF report being asked about ACP by their clinicians. Formulating specific wishes and discussing ACP with a clinician are strongly associated with completing an advance directive. Efforts to improve clinician communication with CF adults around ACP are needed to ensure that discussion of advance directives becomes an integral component of adult CF care.

Introduction

THE SURVIVAL OF PATIENTS WITH CYSTIC FIBROSIS (CF) continues to increase, leading to a continuously growing population of adults living with CF as a chronic illness. In 2004, over 40% of the U.S. CF patient population was over the age of 18.¹ However, CF remains a life-limiting disease, with high mortality secondary to chronic respiratory infections, bronchiectasis, and ultimately respiratory failure. CF presents distinct challenges at the end of life, with tension between the use of preventive therapies, therapeutic options such as lung transplantation, and palliative care. For adult patients with CF, therefore, the discussion of treatment options, including decision making surrounding end-of-life care, is a critical part of their care plan. In this regard, one of the CF

Foundation's goals for improving CF care is the support of all end-of-life care decisions.²

Advance care planning (ACP) is an important tool to promote alignment of the patient's care with his or her needs, goals, and values, and is particularly useful if the patient's ability to make decisions becomes compromised. One purpose of ACP is to develop a treatment plan for those facing divergent options as their disease accelerates. Advance directives such as living wills or designation of a health care proxy provide a legal framework for such planning, and have been increasingly encouraged by many states and health care institutions.³ The American College of Chest Physicians encourages proactive ACP among all patients with chronically progressive pulmonary disease.⁴ Despite these recommendations, studies on ACP in both the general

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and chronically ill patient population have demonstrated a low rate of advance directive completion.⁵

Although ACP discussions are advocated in adult CF care guidelines for all patients regardless of disease severity,⁶ prior research on end-of-life issues in CF have mainly focused on processes and events at the time of death.⁷⁻⁹ Only minimal research exists on the timing of ACP processes in CF, particularly whether these processes are occurring as part of routine care. As a part of the Project on Adult Care in Cystic Fibrosis, we were interested in participants' experiences with processes around ACP. The goals of our study were to assess experiences with advance care planning reported by adults with CF, to assess the reported communication between CF adults, family members, and clinicians regarding ACP, and to identify factors associated with reporting that an advance directive has been completed.

Methods

The Project on Adult Care in Cystic Fibrosis (PAC-CF) is an ongoing prospective, longitudinal panel study of adults with CF 18 years of age or older receiving care at 1 of 10 participating CF centers. The study coordinator at each participating CF center provided data that were used to calculate each individual's predicted probability of surviving 5 years using Liou and coworker's¹⁰ previously published equations. In fall 2004, all adults with a predicted probability of 5-year survival less than 0.975 ($n = 515$) and a randomly selected 25% of adults with a predicted probability of 0.975 or higher ($n = 60$) were approached for inclusion. Compared to adults with CF who were asked to participate in PAC-CF but either refused or could not be contacted ($N = 242$), the 333 adults who enrolled in PAC-CF were more likely to be white, women, and older. Clinically, participants had, on average, better weight-for-age z scores and a higher number of exacerbations. This stratified sampling design was adopted because adults with a very high predicted probability of survival were least informative with respect to the overall PAC-CF goal of examining trends over time in quality of life as CF progresses. That is, adults whose CF was least advanced were sampled at a lower rate because their CF was less likely to progress to moderate or severe during the 3-year time frame of the study. The study protocol was approved by the Institutional Review Boards at Educational Development Center, Inc. and the 10 hospitals in which the participating CF Centers are located.

The data presented here are from the fifth survey of PAC-CF, administered to 303 participants in May 2006. At the time of this survey, 30 of the original PAC-CF participants had either died, received a lung transplant, or were lost to follow-up and were no longer included in the study. This survey included a set of questions about how the respondent would like decisions about their medical care to be made if they became too ill to make decisions for themselves. These questions, which are provided in Appendix A, were developed by the authors and/or adapted from previous surveys.^{12,13} PAC-CF participants were asked to report on their experiences with different components of ACP. Questions were included on how they would like decisions about medical care to be made if they were to become too ill to make decisions on their own. Specifically, we asked whether participants had thought about who should make such decisions,

whether they had any specific wishes, and whether they had heard about or completed advance directive documents such as health care proxies, durable powers of attorney, living wills, or other written instructions. We also assessed how comfortable they were in talking about their decisions with family members and clinicians, and asked about their experiences around communication about ACP issues.

Descriptive statistics, calculated using sample selection weights (the inverse of the probability of selection) to adjust for the disproportionate stratified sampling design, were used to summarize demographic characteristics and to describe experiences with ACP processes. Differences by age, gender, and FEV₁ were assessed using weighted χ^2 test of independence, t test, or analysis of variance (ANOVA), as appropriate. Respondents were divided into four age groups: 18–24, 25–34, 35–44, and 45+ and three groups based on their best FEV₁ (% predicted) as recorded in the 2005 CF Foundation patient registry: less than 40%, 40%–69%, and 70% and greater. We used a multilevel logistic regression model to evaluate possible predictors of reporting the completion of an advance directive.^{14,15} These models, also known as hierarchical linear models,¹⁶ handle the correlated error structures created by the nesting of participants within CF care centers. These hierarchical linear models adjust for potential similarities among participants because they receive care in the same CF center. The dependent variable was a dichotomous indicator of whether an advance directive had been completed. The independent variables included demographic factors (e.g., age, gender, marital status, education), clinical factors, obtained from the CF Foundation Patient Registry (e.g., FEV₁%, exacerbation frequency, colonization with *Burkholderia cepacia*, presence of CF-related diabetes), and factors directly related to the ACP process (e.g., having specific wishes about ACP, having a CF clinician ask about ACP). The multilevel logistic regression model was estimated using SAS PROC NL MIXED, which implements adaptive Gaussian quadrature, a form of numerical integration which has been found to produce statistically reliable estimates for multilevel logistic regression models.¹⁵ All analyses were performed with SAS software version 9.1 (SAS Institute, Cary, NC).

Results

Of 303 surveys, 234 (response rate 77%) surveys were completed. Nonresponders were more likely to be males, younger, and have higher pulmonary function. The mean age ($\pm$ standard deviation [SD]) of respondents was 34 ± 10 years, 61% were female, and their mean FEV₁ (% predicted, $\pm$ SD) was $64\% \pm 23\%$ (Table 1). A majority of the respondents reported current employment or school attendance. In describing their experiences with ACP, 74% (95% CI 68%–80%) of the cohort reported that they had spoken to someone about the medical care they would want if they became too ill to make their own decisions in the future, 65% (95% CI 59%–71%) had thought about whom they would want as their healthcare proxy, and 58% (95% CI 52%–64%) reported having specific wishes about the types of medical treatment they want or would not want if they became too ill to make decisions for themselves in the future. However, only 30% (95% CI 24%–36%) reported completing either a health care proxy, living will or any other type of written in-

TABLE 1. PAC-CF RESPONDENT DEMOGRAPHICS

<i>Summary statistics^a</i>	
Number of respondents (n)	234
Mean age	34 ± 10
Female	61%
Mean FEV ₁ (% predicted)	64 ± 23
One or more exacerbations/year	54%
Have child or children	34%
Are married	51%
Post-high school degree	62%
Are currently employed or are attending school	76%

^aDescriptive statistics were calculated using sample selection weights to adjust for the disproportionate stratified sampling design. PAC-CF, Project on Adult Care in Cystic Fibrosis; FEV₁, force expiratory volume in 1 second.

structions. Table 2 shows the differences in the report of ACP processes based on age, gender, and FEV₁. Respondents ages 18–24 were less likely to report having talked with someone about their care should they become too ill to make decisions, were less likely to report having specific wishes, and were less likely to have reported completing any form of advance directive. Female respondents were more likely to have thought about whom they would want as a health care proxy and were more likely to have completed an advance directive. There were no statistically significant differences in the responses to each of these questions when comparing patients based on lung disease severity as measured by FEV₁.

In reporting on communication with their CF clinicians, 79% (95% CI 74%–84%) of respondents reported feeling comfortable talking to a CF clinician about ACP, and 75% (95% CI 69%–81%) felt comfortable talking to their family about ACP (Table 3). Younger respondents were less likely to report feeling comfortable talking to their family, although they were equally likely to report feeling comfortable talking with CF clinicians. Overall, communication with family members was commonly reported, with a majority (68%, 95% CI 62%–74%) reported having talked with a family member about ACP. A smaller percentage (30%, 95% CI 24%–36%) reported that they had spoken to a friend about such decisions. However, only 13% (95% CI 9%–17%) reported that they have talked with their CF clinician about the care they would want if they became too ill to make decisions for themselves. This was especially true among patients with mild lung disease. In this group ($n = 63$) only 8% (95% CI 1%–15%) reported talking with their clinicians, whereas 28% (95% CI 15%–41%) of those with severe lung disease ($n = 47$) had reported such a conversation. The lack of communication was evident across all types of CF clinicians, with only 12% (95% CI 8%–16%) of the cohort reporting speaking with a physician, 6% (95% CI 3%–9%) with a CF nurse, and 7% (95% CI 4%–10%) with a CF social worker. Only 28% (95% CI 22%–34%) of respondents indicated that their CF clinicians had asked them about ACP, and fewer young adults ($n = 41$, 15%, 95% CI 4%–26%) reported that their clinician had asked them about ACP (Table 3).

We were particularly interested in examining whether clinical or sociodemographic characteristics were associated

with having completed any type of advance care directive. In bivariate analyses, the following characteristics were significantly related ($p < 0.05$) to completion of an advance care directive: female gender (38% versus 25%), age (mean 37 years versus 33 years), having a college degree (41% versus 23%), being married currently or in the past (38% versus 25%), having children (41% versus 28%), having diabetes (48% versus 31%), having specific wishes about treatment decisions (46% versus 15%), and reporting that a clinician had discussed ACP with them (59% versus 24%). In a multilevel hierarchical logistic regression model, the clinical and sociodemographic variables were no longer significantly associated with reporting that an advance directive was completed (Table 4). In this model, which accounts for clustering of respondents at individual CF care centers, only two factors, reporting that a clinician had asked about ACP (OR 4.4, 95% CI 1.5–12.6) and reporting having specific wishes about ACP (OR 7.8, 95% CI 1.9–32.1), were significantly associated with reporting the completion of an advance directive.

Discussion

With the increased survival of CF patients into adulthood, the increased use of intensive care, and the availability of lung transplantation, end-of-life care decisions in CF have become increasingly complex. Prior studies on end-of-life care in CF have focused on events surrounding the immediate time of death; many deaths occur in acute care hospitals,^{7,8,17} and patients are often receiving both therapeutic and palliative interventions at the time of death.⁹ To our

TABLE 2. PROCESSES IN ADVANCE CARE PLANNING AMONG ADULTS WITH CF

	N	% Talk ^a	% Thought ^b	% Wish ^c	% Act ^d
Total	234	74	65	58	30
Age					
18–24	41	56**	62*	43**	10**
25–34	82	72	54	52	25
35–44	71	80	77	64	44
45+	40	89	69	80	37
Gender					
Men	91	67	52**	55	20*
Women	143	78	74	61	36
FEV ₁ %					
<40	47	82	72	72	28
40–70	110	72	58	56	35
>70	63	73	70	56	26

^aHave spoken to someone about the care they would want if they became too ill to make decisions for themselves.

^bHave thought about whom they would like to make their health care proxy.

^cHave specific wishes about the types of medical treatment they would or would not want if they become ill in the future.

^dHave completed either healthcare proxy forms or a living will or other written instructions in which they stipulated the kinds of health care they want or do not want under specific circumstances.

Descriptive statistics were calculated using sample selection weights to adjust for the disproportionate stratified sampling design.

* $p < 0.05$; ** $p < 0.01$.

CF, cystic fibrosis; FEV₁, forced expiratory volume in 1 second.

TABLE 3. ADVANCE CARE PLANNING COMMUNICATION IN ADULTS WITH CF

	N	% Comfortable talking to CF clinician ^a	% Comfortable talking to family	% Talked to CF clinician ^b	% Talked to family	% Clinician asked ^c	% Think clinician should initiate discussion ^d
Total	234	79	75	13	68	28	50
Age							
18–24	41	74	62*	7	45**	15*	54
25–34	82	73	73	8	65	27	46
35–44	71	84	77	19	78	41	49
45+	40	89	93	20	83	24	55
Gender							
Men	91	74	68*	13	59*	25	44
Women	143	82	80	14	73	30	54
FEV ₁ %							
<40	47	83	83	28**	77	43	47
40–70	110	75	72	12	65	31	43
>70	63	83	77	8	66	23	59

^aAre very or somewhat comfortable with talking to their clinician about the medical care they want (or do not want) should they become too ill to make decisions.

^bHave talked to one or more of their CF physicians, nurses or social workers about the kind of care they would like if they became too ill to make decisions.

^cClinician has asked about who they would want to make decisions about their medical care if they become too ill to make their own.

^dThink that their clinician should initiate discussions about medical care they might want if they become too ill to make decisions for themselves.

Descriptive statistics were calculated using sample selection weights to adjust for the disproportionate stratified sampling design.

* $p < 0.05$; ** $p < 0.01$.

CF, cystic fibrosis; FEV₁, forced expiratory volume in 1 second.

knowledge, this is the first systematic report on ACP activities of CF patients prior to the acute end-of-life period.

The results of this survey should put to rest the idea that adults with CF do not want to consider their options for treatment at the end of life. The vast majority of our respondents report having thought about the care they would like if incapacitated, indicating that for many CF adults, ACP begins

well before the end-of-life. Lung disease severity, as measured by FEV₁, was not associated with ACP, highlighting the fact that even patients who are not particularly ill are willing to discuss and act on ACP. Similarly, a large majority of respondents reported feeling comfortable talking about ACP with either CF clinicians or their families, regardless of disease severity.

TABLE 4. FACTORS ASSOCIATED WITH ADULTS WITH CF REPORTING THE COMPLETION OF AN ADVANCE DIRECTIVE^a

Covariate	Estimate	Std error	p	OR (95% CI)
Demographic				
Age	0.03397	0.0251	0.20	
Female gender	1.0322	0.4883	0.06	
Married	−0.0717	0.6581	0.92	
Has children	0.5970	0.4564	0.22	
Lives with a family member	−0.8302	0.8136	0.33	
Has post-high school degree	0.5941	0.5237	0.28	
Clinical				
FEV ₁ 40%–69% predicted	0.6721	0.5720	0.27	
FEV ₁ ≥ 70% predicted	0.8718	0.6881	0.23	
More than one exacerbation in 2005	0.7857	0.5114	0.16	
Colonized with B. cepacia complex	0.9864	0.7195	0.20	
CF-related diabetes	0.7577	0.4866	0.15	
ACP activities				
A CF Clinician has asked about ACP	1.4729	0.4769	0.01	4.4 (1.5–12.6)
Has specific wishes about ACP	2.0532	0.6357	0.009	7.8 (1.9–32.1)
Has thought about ACP	0.2350	0.5710	0.69	
Has talked with someone about ACP	0.4038	0.7385	0.60	
Comfortable talking with family members	1.0453	0.7514	0.19	
Comfortable talking with CF clinicians	0.1709	0.6536	0.80	

^aMultilevel hierarchical model accounts for clustering within CF centers.

CF, cystic fibrosis; FEV₁, forced expiratory volume in 1 second; ACP, advanced care planning.

There is scant literature on the use of ACP among young adults. Small studies have demonstrated that adolescents both with and without chronic illness have developed thoughts about end-of-life care.¹⁸ Culturally, discussing ACP is not part of the experience of many young adults, and so we would expect that completion rates for ACP among young adults would be quite low, although we were unable to find any study which had assessed this. For populations with life-limiting childhood chronic illnesses such as CF we would expect to find higher interest in planning for future care compared to their nonchronically ill peers. However, we found significant differences in the report of ACP activities by age in our cohort as well. Younger CF adults were less likely to have developed wishes, talked about ACP, or completed an advance directive. As young adults, they may be delaying discussions of end-of-life care, particularly with their families, as a sign of hope. This is supported by the lower percentage of young adults who reported feeling comfortable talking family about ACP. In contrast, the majority of young adults reported feeling comfortable discussing ACP with clinicians, yet fewer young adults reported that clinician has spoken to them about ACP and less than 10% had actually talked with a clinician.

The low rate of reporting a completed advance directive by adults with CF is consistent with other studies showing low rates of advance directive completion in the general population,^{3-5,19,20} in patients following lung transplantation,²¹ and in patients with progressive childhood diseases such as Duchenne's muscular dystrophy.²² The low rate highlights the discontinuity between a patient thinking about ACP and completing a document for their medical record. Barriers to the completion of an advance directive may include lack of communication, lack of knowledge, or an unwillingness to discuss end of life care by either clinicians or patients.^{23,24} Our data suggests communication is a significant barrier in CF despite recommendations for adult CF care to incorporate ACP discussions.^{6,8} Most respondents stated that a CF clinician had not spoken to them about ACP, suggesting a communication gap between CF adults and their clinicians around ACP. Our results importantly indicate that patient unwillingness is not a barrier to ACP, as most of our respondents reported talking to family members about ACP and being comfortable talking to clinicians. It is striking that the only factors associated with the completion of an advance directive were whether a patient had developed specific wishes about end-of-life care and whether they had reported that a CF clinician had spoken to them about ACP. Additionally, our analysis controlled for each individual's CF treatment center, and we did not see any significant differences in reported practices between centers. Overall, our results suggest that if clinicians were more active in talking to their patients about ACP, patients would be more likely to complete advance care directives. Successful interventions to increase advance directive completion in other patient populations have advocated direct clinician-patient communication,²⁵⁻²⁷ education of health care providers,²⁸ and discussions with trained nursing or social work staff.²⁹ Similar strategies need to be applied to CF care, particularly by including discussions of advance care planning into routine medical visits.

There is another possibility that might explain the low rate of advance care planning document completion among our adult CF population, namely, that existing ACP docu-

ments do not meet the needs of adults with CF. In the United States, ACP processes were generally designed for those who are older, who were generally healthy during life, and who were stricken with acute disease such as cancer later in life. It may well be that CF, with its relapsing and remitting pattern of disease advancement, the dramatic increases in life expectancy over the past few decades, and the increasingly available option of lung transplantation, is simply not a suitable fit for the existing methods of ACP, such as health care proxies and living wills. The disease trajectory of CF in some ways mimics that of congestive heart failure or adult chronic obstructive pulmonary disease,³⁰ with the important difference of a lifelong experience of illness beginning in childhood. A qualitative study in the UK found that patients and clinicians identified the need for integration of ACP early in the course of disease progression, allowing for co-existence of palliative and therapeutic care over time.³¹ It may well be that more specific advance care planning documents should be designed for those with cystic fibrosis; and yet, our results suggest that few CF adults and clinicians are discussing advance care planning at all. The development of a CF-specific advance care planning document may only partially remediate this lack of communication. In future studies, we plan to examine whether alternate models of advance care decision-making, perhaps those which incorporate discussions about lung transplantation or those which use alternative documents, may be perceived as more useful by adults with cystic fibrosis.

Our study suffers from the limitation of any cross-sectional survey. The data were gathered by self-report and may be subject to over-reporting. We did not examine medical records to see if advance directives were actually completed. We are also unable to assess whether certain changes in health such as a decline in lung function led individual patients to change their thoughts about ACP. Future studies, using qualitative interviews, will examine advance care planning processes and may be able to address some of these limitations.

In summary, we found that although the majority of CF adults report thinking about end-of-life care, developing specific advance care wishes, and communicating with family on such issues, only a minority report completing any documentation supporting their decision. CF adults also report feeling comfortable with ACP discussions, yet very few report being asked about ACP by their clinicians. Finally, we found that formulating specific wishes and reporting that a clinician had asked about ACP are strongly associated with completing an advance care directive. Future studies should particularly focus on understanding and enhancing communication around ACP in CF, so that the worthy goal of the U.S. CF Foundation to support the end-of-life care decision-making of all adults with CF can be realized.

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Appendix A

This section asks questions about how you would like decisions about your medical care to be made if you become too ill to make decisions for yourself.

22. How much have you thought about whom you would like to make decisions about your medical care if you became too ill to make decisions for yourself?

- 1 Very much
- 2 Somewhat
- 3 Not very much

23. Do you have specific wishes about the types of medical treatment you want or don't want if you become very ill in the future?

- 1 Yes
- 2 No
- 3 Not sure

24. How comfortable are you with . . .

	Very Comfortable ▼	Somewhat Comfortable ▼	Not Very Comfortable ▼	Not At All Comfortable ▼	Not Sure ▼
a. Talking with your family about the medical care you want (or don't want) if you become too ill to make decisions for yourself?	1	2	3	4	5
b. Talking with your CF clinicians about the medical care you want (or don't want) if you become too ill to make decisions for yourself?	1	2	3	4	5
c. Appointing someone to make medical decisions for you if you became too ill to make decisions for yourself?	1	2	3	4	5