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Original Article

# Advance care planning in cystic fibrosis: Current practices, challenges, and opportunities☆

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**Abstract**

*Background:* Studies in cystic fibrosis (CF) report late attention to advance care planning (ACP). The purpose of this study was to examine ACP with patients receiving care at US adult CF care programs.
*Methods:* Chart abstraction was used to examine ACP with adults with CF dying from respiratory failure between 2011 and 2013.
*Results:* We reviewed 210 deaths among 67 CF care programs. Median age at death was 29 years (range 18–73). Median $FEV_1$ in the year preceding death was 33% predicted (range 13–100%); 68% had severe lung disease with $FEV_1$ < 40% predicted. ACP was documented for 129 (61%), often during hospitalization (61%). Those with ACP had earlier documentation of treatment preferences, before the last month of life (73% v. 35%; $p$ = <0.01). Advance directives were completed by 93% of those with ACP versus 75% without ($p$ < 0.01); DNR orders and health care proxy designation occurred more often for those with ACP. Patients awaiting lung transplant had similar rates of ACP as those who were not (67% v. 61%; $p$ = 0.55). The frequency of ACP varied significantly among the 29 programs contributing data from four or more deaths.
*Conclusions:* ACP in CF often occurs late in the disease course. Important decisions default to surrogates when opportunities for ACP are missed. Provision of ACP varies significantly among adult CF care programs. Careful evaluation of opportunities to enhance ACP and implementation of recommended approaches may lead to better practices in this important aspect of CF care.


*Keywords:* Advance care planning; End-of-life care; Decision making; Lung transplant

## 1. Introduction

Advance care planning (ACP) is a communication process between patients and health care providers intended to align future medical treatments with goals and wishes of individual patients [1,2]. Tangible outcomes of ACP include advance directives, medical orders for scope of treatment, and appropriate documentation and communication of treatment preferences.

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Cystic fibrosis (CF) is a genetic disease with limited life expectancy due to progressive impairment of lung function. Respiratory failure is the most common cause of death [3]. The few published studies addressing ACP in CF suggest that it often occurs late in the illness course, if at all [4–6]. In one study, only one-third of adults with CF reported having been asked about ACP by a health care provider, and a similar proportion completed an advance directive [7]. Single center investigations have revealed late or absent ACP and advance directives, "do not resuscitate" (DNR) orders written at the very end of life, and a majority of deaths occurring in the hospital with intensive treatments ongoing until the time of death [4,6]. While ACP is recommended for people with CF [8–11], there are currently no formal guidelines for ACP in this population.

In the US, most CF care is provided in care centers accredited by the Cystic Fibrosis Foundation (CFF). Multidisciplinary care teams, including physicians, nurses, social workers, dieticians, physical therapists, and respiratory therapists, provide coordinated CF care. While only physicians and licensed practitioners may formally enter orders for limitations on life-sustaining treatments that follow from ACP conversations, patients may choose to have conversations about treatment preferences and future medical care with any trusted provider within the care center. Much of CF care in the US is provided under direction of guidelines [12], but the personal, patient-centered nature of treatment decision making and lack of formal guidelines for ACP as part of CF care suggests there may be variation in practice for ACP with patients among CF care centers.

Previous studies and clinical experience highlight challenges to ACP, including some that are relatively unique to CF. While shortened life expectancy [3] might seem to trigger ACP, variable disease progression makes prognostication difficult. Lung transplant, an option for select patients with advanced disease, could delay ACP [13]. Family caregivers often participate in medical decision making; thus, planning may not reflect an individual's preferences and important decisions may be deferred to surrogate decision makers when lung disease progresses more rapidly than anticipated [5,14,15]. Several multidisciplinary CF care team members may participate in ACP, but lack of clarity on the responsible party may delay initiation of the process. Additionally, while practice standards exist for many aspects of CF care, there is lack of consensus about appropriate timing of ACP. To our knowledge, no national study of end-of-life care in CF has been performed in the United States. We undertook this study to examine practices for ACP in US-based adult CF care programs, evaluating these and other potential challenges and informing opportunities for improvement.

## 2. Methods

We adapted a web-based chart abstraction tool from the End-of-Life Chart Review Tool [16] to collect information about patients who died from complications of CF, including age, cause, and location of death, conversations about ACP, and advance directives. The tool was pilot tested at the authors' institutions and further refined (Appendix 1).

We invited all 113 currently accredited US adult CF care programs to complete a review of the last five CF deaths at their institutions occurring from 2011 to 2013. One program reported six deaths, and all were included; many programs had less than five deaths occur during that time period. Approval for the study was obtained from the Institutional Review Board at each participating institution. Programs were offered a $100 stipend by the CFF for each abstraction. A pre-programmed query in the CFF Patient Registry [17] was used to identify patients who died within the designated time frame. Patients under age 18 years were excluded. Patients dying after lung transplantation (approximately 16% of CF deaths in the US in this time period(12)) were excluded as their care differs significantly after the procedure, and many of these patients have less consistent follow-up with multidisciplinary CF care teams after transplant. Data from patients with non-respiratory causes of death were excluded (14 unknown, 7 cirrhosis, 17 unrelated to CF) because their clinical courses were not likely representative of those of patients dying from progressive lung disease.

Two researchers (JG and RIC) independently reviewed data and made recommendations regarding potential data errors, recoding of data entries, and exclusions from analysis. Discrepancies were reviewed with the research team and final decisions were made by consensus. For purposes of analysis, we defined ACP as "documented conversations about treatment preferences between patients and CF health care providers" and assessed for tangible outcomes of ACP include advance directives, medical orders for scope of treatment, and appropriate documentation and communication of treatment preferences. Conversations between surrogate decision makers and health care providers were included in the analysis as "conversations about treatment preferences" but were not considered ACP even if orders for limitations on life-sustaining treatments were documented as these decisions can be made by surrogates when patients are no longer able to communicate with health care providers.

Summary statistics were used to describe results. Chi-square tests were used to determine differences between groups, such as those with and without ACP. A multivariable logistic model was used to determine whether demographic characteristics (gender, age, lung function, insurance, and transplant status) and ACP measures (location of ACP, advance directives, lung transplant, and palliative care consults) were predictive of outcomes of timing of ACP and timing of advance directives. Variables having a probability of $<0.20$ from bivariate correlations and a variance inflation factor $<2.5$ ($\geq 2.5$ indicates too many correlations with other variables in the model) were entered into the model using a web-based logistic regression building tool [18]. Missing or "not applicable" responses were excluded from the analysis.

A funnel plot was used to determine whether ACP differed among CF care programs. The ACP rate for each program reporting four or more patient deaths was plotted with ACP on the *y*-axis and number of deaths on the *x*-axis. The funnel plot is a statistical process control method that uses 3-sigma control limits ($p = 0.001$) to determine outliers and uses the Wilson method to account for the small number of deaths per program