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Original Article

End-of-life practice patterns at U.S. adult cystic fibrosis care centers: A national retrospective chart review

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Abstract

Background: There are many challenges to providing end-of-life care (EOLC) to people with cystic fibrosis (CF).
Methods: Chart abstraction was used to examine EOLC in adults with CF who died between 2011 and 2013.
Results: We reviewed 248 deaths from 71 CF care centers. Median age at death was 29 years (range 18–73). While median FEV1 was in the severe lung disease category (FEV1 < 40%), 38% had mild or moderate lung disease in the year preceding death. The most common location of death was the intensive care unit (ICU, 39%), and 12% of decedents were listed for lung transplant. Fewer of those dying in the ICU personally participated in advance care planning or utilized hospice or Palliative Care Services ($p < 0.05$).
Conclusions: Adults dying with CF in the United States most commonly die in an ICU, with limited and variable use of hospice and Palliative Care Services. Palliative care and advance care planning are recommended as a routine part of CF care.

Keywords: End-of-life care; Hospice; Palliative care; Advance care planning; Lung transplant; Cystic fibrosis

1. Introduction

Cystic fibrosis (CF) is evolving from a life-limiting childhood disease to a chronic disease of both childhood and adulthood. Median survival in CF has increased from 14 years of age in 1969 [1] to over 40 years in 2015 [2]. An increasing number of people with CF now survive into or beyond their 50s [2], and infants born with CF now may expect to survive into their 50s and beyond [3]. Previous studies indicate that many adults with CF receive intensive care up to the final days and weeks of life including hospitalization, mechanical ventilation, intravenous antibiotics and routine CF medications [4,5]. In addition, referrals to hospice and palliative care (PC) are limited or late in the course of disease [6,7].

Palliative care includes the prevention and relief of suffering by means of early identification and management of physical, psychosocial, and spiritual problems [8]. It should be available throughout the course of a chronic serious illness such as CF, concurrently with restorative treatment [9,10]. End-of-life care (EOLC) is one component in the continuum of PC, focusing on the last days, weeks, or months of life [11] with a greater focus on

Abbreviations: ICU, intensive care unit; PC, palliative care; EOLC, end-of-life care.

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comfort measures and a shift in goals of care from extending quantity of life to improving quality of life. Because of the physical and emotional symptom burden of CF [12–19], PC can significantly improve quality of life for people with CF, especially as the disease progresses and at the end of life. Historically, CF EOLC did not include the use of life-sustaining treatments such as mechanical ventilation since this type of care was felt only to prolong the dying process [20]. Now, in the era of lung transplantation and advancing survival, life sustaining interventions may be offered even to patients with advanced CF lung disease, often with hopes of bridging to lung transplant [21–24]. If transplant or reversal of organ failure is not possible, patients may die while on or soon after withdrawal of life sustaining therapies. Therefore, simultaneously planning for lung transplant and end of life is necessary at this stage of illness.

Providing optimal EOLC for people with CF remains challenging. Variable disease progression makes prognostication difficult [5]. Prognostic uncertainty and pursuit of lung transplant could delay discussions about EOLC and advance care planning (ACP) [25,26]. Referral to hospice may be challenging since many CF treatments that can improve quality of life may be cost-prohibitive for patients enrolled in hospice care. There is hesitance to treat pain and symptoms with opioids and benzodiazepines, due to fear of addiction, dependence, respiratory depression, constipation, and impaired airway clearance due to cough suppression [16,18].

While the body of literature for PC and EOLC in CF is growing, no large multi-center research study has evaluated CF-specific EOLC practices in the United States. A previously published article on ACP practices in CF care [26] from the same retrospective chart review uncovered variability in ACP practices among adult CF programs, and many opportunities to enhance this important aspect of care. The goal of this study was to review current EOLC practice patterns among U.S. programs caring for adults with CF to understand issues faced by people with CF as lung disease advances, to examine how practices vary across centers, and to inform efforts to improve PC and EOLC.

2. Methods

Development and administration of the web-based chart abstraction tool was previously described [26]. The End-of-Life Chart Review Tool [27], a publicly available comprehensive tool, was adapted by the research team to collect information about people who died with CF, including age, cause, and location of death, advance directives, and symptoms. Information specific to CF including lung function and lung transplant referrals were added by the research team. The tool was pilot tested at the authors' institutions and further refined.

All 113 US adult CF care programs accredited at the outset of the study were invited to complete a review of the last five CF deaths at their institutions occurring between 2011 and 2013. Adults 18 years or older were included, along with all causes of death. CF care programs were instructed not to include individuals with CF dying after lung transplantation (approximately 19% of CF deaths (2)) as many patients transfer their primary site of care from the multidisciplinary CF care center to the lung transplant center. Approval for the study was obtained from the Institutional Review Board at each participating institution. Programs were offered a $100 stipend for each chart abstraction.

Two researchers 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 made by consensus.

Summary statistics were used to describe results. Chi-square tests were used to determine differences between groups. A p-value of 0.05 was used to determine statistical significance. A funnel plot was used to determine whether the percent of patients receiving services prior to death, such as palliative care, and location of death, such as home, differed among CF care programs. The funnel plot is a statistical process control method that uses a 3-sigma control limits (p-value 0.001) to determine outliers and used the Wilson method to account for small numbers of deaths [28]. Only programs reporting four or more patient deaths were compared.

3. Results

Information from 254 deaths was submitted by 72 adult CF care programs (63% of accredited US programs). Six patients were excluded because they died after lung transplant or outside of the designated time frame. Data from the remaining 248 patients from 71 programs were analyzed. Two programs reported six deaths instead of the recommended five, and all were included; 35 programs had less than five reported deaths during that period. Patient demographic information is displayed in Table 1. Median age at death was 29 years (interquartile range 24–38) and 54% were female. Median highest and lowest FEV1 were both in the severe lung disease category (FEV1 < 40% predicted) [10] in the year preceding death, and 38% had mild or moderate lung disease based on their highest FEV1. Thirty patients (12%) were on the lung transplant wait list at the time of death. The most common type of insurance was Medicaid (36%), followed by private insurance (34%), and Medicare (26%). For analysis, those with private insurance or Medicare were considered fully insured, and those with Medicaid or no insurance were considered underinsured. The most common primary cause of death was respiratory or cardiac failure (n = 210, 85%), followed by liver failure (n = 7, 3%). Suicide and overdose together made up an additional 10 deaths (4%). Palliative Care or Hospice services were utilized by 124 patients (50%), including those who were documented to have hospice alone (n = 22), PC alone (n = 47), or both concurrently (n = 55). Fig. 1 shows that the intensive care unit (ICU) was the most common location of death (n = 96, 39%), with the majority of those using mechanical ventilation within the last week before death.

Information about individuals with CF who died in the ICU compared with those who died elsewhere is shown in Table 2. Fewer of those dying in the ICU personally participated in ACP (rather than relying on surrogate decision makers) compared with those dying elsewhere (p = 0.006), and fewer utilized hospice or Palliative Care Services (p < 0.001). Five specific

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