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Family Caregiver Perspectives on Symptoms and Treatments for Patients Dying From Complications of Cystic Fibrosis

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Abstract

Context—Most patients with advanced cystic fibrosis (CF) die from respiratory failure and experience distressing symptoms as lung disease progresses. Little has been reported about symptom management and the continuation of disease-specific treatments near the end of life for patients with CF.

Objective—We aimed to describe symptom prevalence, symptom management, and frequency of use of disease-specific treatments for patients dying from complications of CF.

Methods—We conducted semistructured interviews about end-of-life care in CF with bereaved family caregivers and asked questions about symptoms and treatments.

Results—Twenty-seven caregivers answered questions about symptoms and treatments. Caregivers reported that distressing symptoms were common during the last week of life, including dyspnea (100%), fatigue (96%), anorexia (85%), anxiety (74%), pain (67%), and cough (56%). Most caregivers felt that symptom control was "somewhat good." Many reported that

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medical providers “did the best they could” to manage symptoms but four (15%) recalled no physician inquiry about symptoms. Caregivers expressed beliefs that symptoms could not be controlled and described concerns about side effects and potential for hastening death with the use of opioids and anxiolytics. Patients received numerous disease-specific treatments, and caregivers described many of them as uncomfortable but necessary.

Conclusions—Distressing symptoms are common in dying CF patients, and disease-specific treatments also cause discomfort. Many family caregivers have low expectations for symptom control. This exploratory research can be used to inform clinical interventions to improve symptom management for patients with advanced CF. J Pain Symptom Manage 2010;40:829–837.

Keywords

Cystic fibrosis; symptom burden; symptom prevalence; palliative treatments

Introduction

Cystic fibrosis (CF) is a lifespan-limiting genetic disease with a predicted median survival in the United States for 37.4 years.[1] The natural history of CF is a progressive decline in lung function secondary to chronic airways infection, with death usually resulting from respiratory failure.[1,2] Lung transplant for advanced CF lung disease is a therapeutic option that may improve survival and quality of life for selected patients.[3,4] Current five-year survival after lung transplant for CF is about 50% in the United States,[5] and bronchiolitis obliterans syndrome (BOS), another chronic progressive lung disease, which is felt to be a manifestation of chronic rejection, is the most common cause of death in lung transplant recipients surviving longer than one year after transplant.

Distressing symptoms, such as dyspnea, cough, pain, and fatigue, are common in patients with CF[6-11] and are known to increase as lung disease progresses.[12-14] BOS symptoms and treatments often mirror those of CF.[15] Although most patients with CF die from respiratory failure because of primary disease complications or BOS, little research defines the impact of symptoms on the end-of-life experience of these patients. Despite evidence that medications for dyspnea and pain can be used safely,[16,17] concerns about respiratory depression in patients with advanced CF lung disease have been described[13,18-20] and may act as barriers to effective palliation.

As CF lung disease and BOS progress, the frequency and severity of respiratory exacerbations tend to increase, resulting in both increased symptoms and intensification of treatments, including oxygen and other respiratory support, airway clearance therapies, aerosolized medications, oral and intravenous antibiotics, and nutritional supplements given orally or via feeding tube. Our goal for this study was to use family caregiver response to interview questions about end-of-life care in CF to better characterize symptom prevalence, symptom assessment and management, frequency of use of disease-specific treatments, and attitudes about treatments used for patients with CF dying from advanced lung disease or progressive BOS after lung transplant.

Methods

Subjects

We used our institutional CF patient registry to identify all patients primarily followed at our center who died from respiratory failure because of CF or BOS between 1996 and 2006 (n = 54 patients). Current contact information was found for the primary contacts listed for 45 of these patients. We mailed letters to them requesting their participation in a study about end-

of-life care and provided an opt-out letter. Of the 45 eligible caregivers, 39 answered telephone calls, and 33 (73% overall), corresponding to 33 individual patients, gave consent for participation. Of six who declined, two cited sadness related to the patient's death, one discomfort because of the declining health of another family member with CF, and three provided no explanation. Six patients treated with mechanical ventilation during the entire last week of life were too heavily sedated for caregivers to be able to answer questions about symptoms, leaving 27 caregivers to complete this portion of the interview.

Interviews

Each caregiver completed a semistructured interview about the experiences of corresponding patients (27 caregiver-patient pairs). This qualitative approach was chosen because of the paucity of data about end-of-life experiences, including symptoms and symptom management in advanced CF, a small potential subject pool, and our goal of exploring concepts that might inform the development of future studies of symptom management in CF. We adapted questions about end-of-life care, including symptom assessment and management, from existing questionnaires.[21-23] We asked closed-ended questions about the presence or absence of common symptoms experienced by patients with advanced lung disease because of CF and BOS, including dyspnea, fatigue, anxiety, pain, cough, anorexia, and nausea, and asked caregivers to identify the most distressing symptom from this list. We also asked scaled questions about the frequency of symptom assessment and treatment by medical providers, frequency of disease-specific treatments, and perceived effectiveness of and burden of these treatments. Caregivers were given the opportunity to expand on answers to all closed-ended questions.

The interview script was reviewed by all investigators, the study was approved by the University of North Carolina Institutional Review Board, and then pilot tested with three caregivers (two parents and one sibling) of patients who died from 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, with an average length of 55 minutes (range 30–88 minutes).

Medical Record Review

Systematic review of medical records was used to verify caregiver reports of demographic information, lung function, lung transplant status, medications and treatments prescribed during the last six months of life and during the last week of life (for patients who died in the hospital), and location of death.

Data Analysis

Quantitative data analysis was performed using Stata version 9 software (Statacorp, College Station, TX). Summary statistics were used to analyze data extracted from medical records, categorical questions about the presence of specific symptoms and administration of treatments, and scaled questions about symptom assessment and perceived effectiveness of treatments. 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.

Interview transcripts were reviewed, and qualitative content analysis with an inductive approach was used to delineate predominant themes from expanded responses to closedended questions.[24] These themes served as codes, which were then applied to remaining transcripts independently by two investigators (M. S., and K. N.) using MAXQDA2007 (©VERBI Software, Marburg, Germany). Codes were refined, and additional codes were developed as the analysis progressed. Differences in coding were

reconciled by consensus. Representative quotes were selected for inclusion in presentation of the data.

Results

Characteristics of Caregivers and Their Corresponding Patients

Caregiver characteristics are summarized in Table 1. Most were parents and identified themselves as primary caregivers. Nonparticipants did not differ significantly from participants with regard to age of the patient at death (22.7 years for nonparticipants vs. 24 years for participants, $P = 0.90$), relationship to the patient (84% of nonparticipants were parents vs. 82% of participants; $P = 0.67$), or location of the patient's death (100% in hospital for nonparticipants vs. 81% in hospital for nonparticipants; $P = 0.24$).

Characteristics of corresponding patients also are summarized in Table 1. Most had severe lung disease, with 59% having an average forced expiratory volume in one second (FEV_1) below 30% of predicted over the six months preceding death, a cutoff that commonly prompts consideration of referral for lung transplantation. Patients were overall quite debilitated and used numerous disease-specific treatments during the six months preceding death and in the final week of life (Fig. 1). All patients died from respiratory failure related to either advanced CF lung disease ($n = 14$) or BOS after lung transplantation for CF ($n = 13$). Average survival after transplant was 4.1 years (range 1.2–10.2 years). Interviews were conducted on an average of 7.5 years (range 1.8–10.9 years) after patient death.

Themes elicited from caregiver responses to questions about symptoms and treatments are presented below and are summarized in Table 2.

Symptoms Experienced by Patients with Advanced CF Lung Disease

As shown in Fig. 2, dyspnea, fatigue, and anorexia were the most commonly reported symptoms in the last week of life, experienced by at least 80% of the patients. Over half of the patients also were reported to have anxiety and pain. There was no difference in frequency or nature of symptoms in patients dying from CF vs. BOS.

Dyspnea was reported to be the most distressing symptom for most patients (52%). Typical caregiver descriptions of dyspnea included, "She was having problems breathing all the time" and "The shortness of breath was horrible. If I had to describe it, I would say she practically smothered to death."

Anxiety was another common and distressing symptom, with nearly 20% of the caregivers reporting that patients had frank panic attacks. Caregivers often alluded to the overlap between dyspnea and anxiety. One stated, "[Patient] was very, very, very agitated, and just couldn't breathe and was very frustrated.she's just agitated and just nervous, tense, just real upset about everything. I think the anxiety was the worst symptom." Another described the intersection of these symptoms by stating, "I think [patient] felt like he wasn't getting any air. It was like an anxiety attack but he would be registering 97 or 98 [percent oxygen saturation]." In the context of describing a patient's dyspnea, another caregiver said, "You can understand her anxiety."

With regard to other symptoms, pain was most often described as localized to the chest, but headache and joint pains were also common. Although fatigue was identified by most caregivers, few remarked specifically on its impact on patient functioning. Anorexia and nausea were also common symptoms, with eating often being described as laborious: "[Patient had] no appetite. None. I would have to beg. In fact, she would eat a little bit and drink her [nutritional supplement] really just to shutme up because I, well I knew she

couldn't survive without it." Although many patients received tube feedings, few caregivers discussed patient tolerance of this form of nutrition.

Symptom Assessment

When asked about how often physicians asked about distressing symptoms, none of the caregivers felt that inquiries were excessive. A majority (55%) felt the frequency of inquiries was "just right," with one caregiver stating, ".they would come in to say how is [patient], is she uncomfortable and things like, she's okay and she's not in any distress, and, they were just very attentive." However, 27% felt such inquiry occurred "not often enough," and 15% did not recall physicians ever asking about distressing symptoms. Symptom assessment for hospitalized patients was commonly described as part of nursing care, with nurses relaying this information to physicians; thus, direct patient-physician communication about symptoms did not always occur. One caregiver recalled, "[Physicians] didn't ask us. They would ask the nurses."

For some patients, limitations on symptom assessment were imposed by escalation of care to include mechanical ventilation and the associated need for sedation. One caregiver remarked, "I think she was comfortable. I mean she wasn't… thrashing around or you know acting like she was in any distress." Another said, "It seemed like sometimes when the blood pressure would go up or the heart rate would get a little stronger, the nurse may come in and say that she could be in a little bit of pain, but they would try to assure me that she was not in any pain."

Symptom Management

Only 9% of the caregivers reported that overall symptom control was "very good," with most (71%) describing it as "somewhat good." The remaining 20% felt symptom control was "somewhat poor" or "very poor." The predominant theme that emerged concerning symptom control was that medical providers "did the best they could" to manage distressing symptoms. One caregiver said, "For the condition she was in, I think [symptom control] was good. I don't believe they could have done anything more." Others noted perceived limitations of medical treatments, stating, ". they controlled what they could with the shortness of breath. Of course you know there was not a lot they could do at that point," and "[Symptoms] couldn'tbe completely controlled. There were not adequate medications." Another commented on delays in receiving medications prescribed to alleviate symptoms: "[Symptoms] were controlled pretty well given the system. The system is just slow. There is a long time in between feeling the pain and getting something to relieve it."

Another pervasive theme was the difficulty of managing distressing symptoms because of the discomfort caused by disease-specific treatments. Chest physiotherapy, which facilitates clearance of lower respiratory secretions, was most often identified as the most burdensome treatment, primarily because it may be painful. Bilevel positive airway pressure (BiPAP), also felt to cause discomfort, was described by one caregiver as follows: "And she could not tolerate the BiPAP. The pressure of it gave her terrible headaches. It was the only time that she said I can't stand that, I can't do that." Many caregivers described patients feeling there was no choice but to endure these associated discomforts, with one stating, ".they upped her oxygen, they put her on a face mask. That was hard for her because she didn't want anything on her face, you know. But then she realized that if it helped that she could breathe better. And then she did allow them to do the face mask."

Attitudes About Medications Used to Treat Symptoms

Although all patients ultimately treated with mechanical ventilation received both opioids and benzodiazepines, according to caregiver reports, only 55% of the patients not treated

with mechanical ventilation received opioids and 37% received anxiolytics during the last week of life. Although we did not specifically inquire about previous use of these medications, only two caregivers mentioned chronic use of anxiolytics, and none mentioned chronic opioid use during the last six months of life. Review of medical records substantiated these reports. Indications for opioids identified by caregivers were pain and dyspnea, and indications for anxiolytics were anxiety and dyspnea. A predominant theme emerging from caregiver responses was reduction in distressing symptoms with the use of medications. Comments about medication effects included, "… when it would get to the point where she would kind of panic, so then they gave her [diazepam] to calm her down. You know that worked really well…" and "They had her on a morphine pump to help keep her comfortable, and later on she had inhaled morphine. The nurse said it wouldn't make her lungs better but it did make her feel like she was breathing again." Although increased fatigue and drowsiness with administration of opioids and benzodiazepines were identified by some caregivers, symptom relief also was acknowledged: "And so the pain meds were increased and one day he'd be talking and then the next day he'd be out of it, but he was more comfortable and he was more at peace."

Another theme emerging from caregiver responses was concern about medication side effects, including respiratory depression, drowsiness, confusion, and fatigue. One caregiver felt effective treatment of pain was limited by side effects, noting, "I think it was controlled as much as they could without putting her out." Other caregivers reported patient and caregiver hesitance in accepting certain medications, with one stating, "[Patient] had to read up on morphine before she would [use it]. She had to read up on it and I remember what she said, she said it can cause my breathing to slow down. I said yes it can. I said it can even cause it to stop." Some medical providers tried to ease these concerns. According to one caregiver, "[Nurse] said if you think she needs medication for chest pain, and you can't push the button because you're afraid it will interfere with her breathing, he said, you call me and I will push it."

A final theme was attachment of stigma to certain medications by caregivers, patients, and medical providers. Some medical providers alluded to certain medications being appropriate only immediately before death or even hastening death. One caregiver described, ".we finally got to the point [death was imminent] and [medical team] was really good about explaining the pain meds and what they could do and you know that she wouldn't be able to respond once they upped the meds and it was like, let's make her comfortable because that's what's more important. Things will speed up once we do this and that's why it's time. She's ready." Another said, "…you know when they start putting you on morphine and stuff it's only a matter of time." Others referred to issues with medication misuse or abuse, making comments like, "I think he wanted to be kept sedated so he asked us several times to push the [lorazepam] and the morphine. I mean he said well don't be stingy with them ." and "[the physician] said well, if you want to make her a dope addict or just keep her doped up, I mean it was just so. I said, look I didn't make these decisions. I just want to make her comfortable."

Discussion

We interviewed caregivers of patients with CF who died from respiratory failure because of CF lung disease or post-lung transplant BOS about the symptoms that patients experienced and treatments they received near the end of life. Caregivers reported a high prevalence of many distressing symptoms, including dyspnea, fatigue, anorexia, anxiety, pain, and cough. Symptoms experienced by patients dying from CF lung disease and BOS did not differ. Many caregivers had low expectations for symptom control, expressing beliefs that symptoms simply cannot be controlled and that potential side effects of medications for

distressing symptoms limit treatment options. Disease-specific treatments such as chest physiotherapy, BiPAP, enteral nutrition, and standard medications were commonly administered near the end of life and were often perceived by caregivers to cause distressing symptoms. These findings may have implications for the development of interventions to improve symptom control and selection of disease-specific treatments for patients with CF dying from advanced CF lung disease and from BOS after lung transplant.

Symptom management in CF patients with advanced lung disease is complex. Symptoms may be due both to the underlying disease process and to side effects of treatments, and overlap in symptoms often occurs. Despite a call for careful attention to and skilled management of symptoms that may cause suffering in patients with CF,[14,16] our findings suggest that improvements are needed. Many barriers to symptom management have been identified in other life-limiting illnesses. These may include inadequate assessment and/or communication about symptom presence and impact on function and quality of life, insufficient training of medical providers in prescribing treatments, and concern about side effects of certain medications and treatments.[21,25,26] A common finding in our study was that physicians often obtained information about dying patients' symptoms indirectly from nurses rather than asking patients or caregivers directly. Studies addressing provider communication skills as well as perceptions and attitudes about symptom management could inform interventions to improve symptom management for dying CF patients. Additionally, studies addressing how to reduce and counteract side effects and discomforts may lead to improvement in tolerance of disease-specific treatments, which providers, caregivers, and patients alike feel are necessary in the management of advanced lung disease because of CF or BOS.

This study is exploratory and should be interpreted in light of some limitations. First, this single-center study does not necessarily reflect practices at other institutions. The interview script was used for a larger study of end-of-life care in CF, and questions about symptoms and treatments were developed to broadly explore symptom assessment and management, not to address the impact or management of specific symptoms. Because of the study design, patients themselves did not report symptoms, but rather we asked caregivers about these issues as experienced near death. Because caregivers are closely involved in the care of many patients with CF throughout the lifespan,[27] their perspectives are useful in addressing the study questions. We asked these caregivers to recall details of discussions that took place many years before the interview. Recall of terminal illness by relatives has been reported in the shorter term, [28] and it is reported that proxies' perceptions of patient' symptoms are less reliable than for other aspects of quality of care at the end of life,[29] but bereavement studies yielding reliable data about symptoms have been conducted up to 10 years after death.[21,30] Because of concerns about recall bias, we compared recalled events to factual demographic information and disease-specific information previously abstracted from medical records and found concordance.

Despite these limitations, we feel our findings highlight deficiencies in the care of complex, chronically ill patients with CF bothbefore and after lung transplant, and identify areas for further research in symptom assessment and management for these patients. A final caregiver quote highlights both an expectation and an opportunity: "But I think today, today, definitely I think they could [treat symptoms] much better than they could back then. I think palliative care is much more advanced and perhaps there are medications and procedures that are more finely tuned." Defining barriers to symptom assessment and management and then designing interventions to overcome these barriers is one potential approach.

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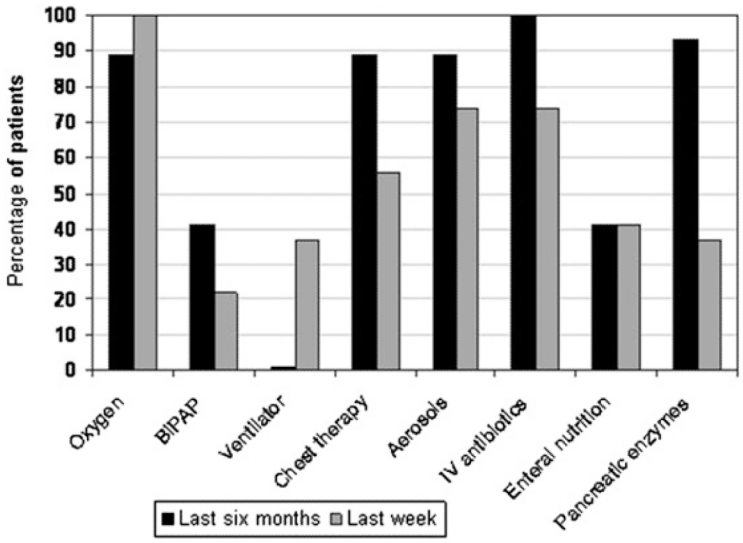

Fig. 1.
Disease-specific treatments used during last six months and last week of life.

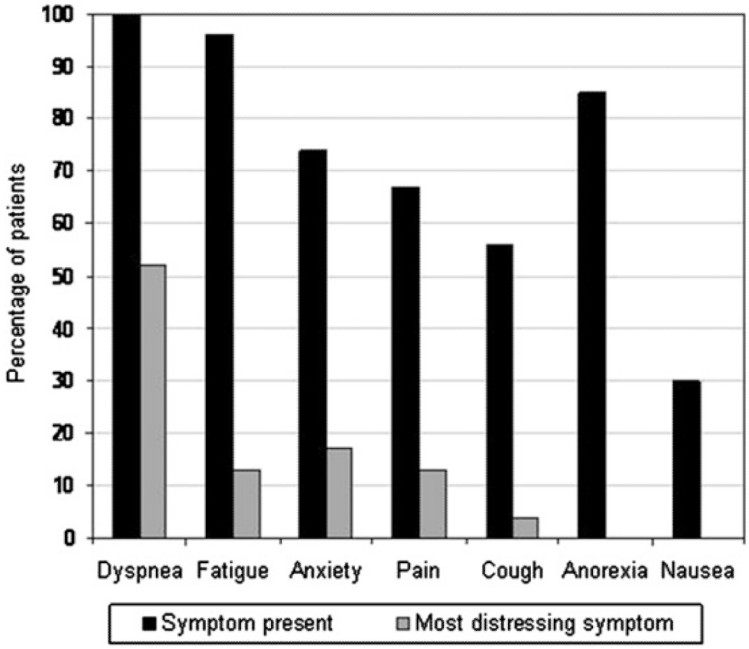

Fig. 2.
Prevalence of symptoms and most distressing symptoms as reported by caregivers.

Table 1

Characteristics of Caregivers Participating in Interviews and Their Corresponding Patients (*n* = 27 Caregiver-Patient Pairs)

Caregiver and Patient Characteristics	Number (range or %)
Caregivers	
Relationship to patient	
Parent	24 (88)
Grandparent	1 (4)
Spouse	1 (4)
Friend	1 (4)
Mean age in years (range)	58 (32–82)
Female gender	23 (85)
Primary caregiver to patient	26 (96)
Patients	
Mean age at death in years (range)	24 (8–47)
Female gender	18 (67)
Lung function: average FEV_1 over last	
6 months of life	
Less than 40% of predicted[a]	24 (89)
Less than 30% of predicted[b]	16 (59)
Lung transplant status	
Underwent transplant	13 (48)
Awaiting transplant	5 (19)
Declined transplant	2 (7)
Not referred for transplant[c]	7 (26)
Location of death	
Hospital, intensive care unit	9 (33)
Hospital, medical ward	13 (48)
Home	5 (19)
Functional status during last 6 months of life	
Very limited	9 (33)
Somewhat limited	14 (52)
Not very limited	4 (15)
Not limited at all	0 (—)

[a] Severe lung disease in CF is defined as FEV_1 below 40% of predicted.

[b] FEV_1 below 30% is the cutoff below which lung transplantation may be considered.

[c] Of those not referred for transplant, five did not meet medical criteria, one had medical contraindications to transplant, and one was nonadherent to medical therapies.

Table 2

Themes Elicited From Analysis of Caregiver Responses to Questions About Symptoms and Treatments

Topic	Themes
Symptoms experienced by patients with advanced CF lung disease	• Individual patients have numerous symptoms • Symptom overlap is common • Many disease-specific treatments cause discomfort but are felt to be necessary
Symptom assessment	• Nurses, not physicians, provide primary symptom assessment • Use of mechanical ventilation limits symptom assessment
Symptom management	• Medical providers do the best they can to treat symptoms • Medical treatments have limitations; some symptoms cannot be controlled • Treatment side effects limit optimal symptom management
Attitudes about medications used to treat symptoms	• Medications may reduce distressing symptoms • Concern about medication side effects • Attachment of stigma to certain medications by patients, caregivers, and medical providers