PALLIATIVE CARE NETWORK OF WISCONSIN

FAST FACTS AND CONCEPTS #265
PALLIATIVE CARE FOR PATIENTS WITH CYSTIC FIBROSIS

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Background Cystic fibrosis (CF) is an autosomal recessive disorder of the cystic fibrosis transmembrane regulator (CFTR) that affects the respiratory tract, pancreas, intestines, male genital tract, hepatobiliary system, and exocrine sweat glands resulting in multisystem disease. This *Fast Fact* discusses the natural history of CF and issues related to palliative care for patients with CF.

Morbidity and Death in CF The life expectancy of patients with CF has improved over the last 50 years. In 1959, the median age at death was 6 months and in 2008 it increased to 27 years. For those born in the year 2010, the median age of survival is predicted to be greater than 50 years (1). This improved survival is attributed to advances in the supportive care of patients including early diagnosis, family support, attention to nutrition, infection control, and the use of nebulised mucolytics and antimicrobials (3).

- Chronic progressive pulmonary disease and respiratory failure remain the major cause of morbidity and mortality. End-stage lung disease is characterized by cysts, abscesses, and fibrosis of lungs and airways. Patients frequently die from overwhelming lung infections.
- Factors associated with earlier death in CF include the deltaF508 mutation, pancreatic insufficiency, low body mass index, certain infections (*Pseudomonas aeruginosa*, *Staphylococcus aureus,* and *Burkholderia cepacia*), low socioeconomic status, tobacco smoke exposure and female sex (2,3).
- For select candidates, lung transplantation improves survival and quality-of-life. The 5-year survival post-transplant is about 50%.
- Other important causes of morbidity include CF-related diabetes mellitus, pancreatic insufficiency (manifesting as fat malabsorption, poor growth, and hemolytic anemia), hepatobiliary disease (including portal hypertension and varices), male infertility, and meconium ileus in newborns.
- CF patients commonly suffer from cough, fatigue, and dyspnea; these symptoms tend to worsen as lung disease progresses. Chest wall and back pain from cough, pleurisy, rib fractures, and chest physiotherapy is common (4). Fears of addiction, respiratory depression, and concern that chronic opioid use complicates lung transplant success can create barriers to appropriate opioid administration (3,5,6).
- With multiple medications, therapies, and nutritional supplements required each day, routine care can be physically and emotionally exhausting for CF patients and family members.
- Adolescents with CF report a high quality-of-life and they abuse substances less frequently than their healthy counterparts (7). Nonetheless, in this age group, chronic anxiety and depressive disorders are common and treatment adherence routinely declines (7).

Common Issues Near the End-of-Life

- Prediction of short-term mortality is difficult and largely subjective. Most predictive models focus on clinical severity of pulmonary disease using age, FEV1%, BMI, the presence of pancreatic insufficiency, and opportunistic infections as parameters. These models however are designed to stratify lung transplant candidates rather than assist with end-of-life care planning (3,9).
- The dynamic, chronic, progressive course of CF creates challenges for timing goals-of-care discussions. Periodic planned meetings amongst patient, family, and care team are advised. Single-center studies reinforce that, despite documented need, palliative care consultation is rarely offered until days before death or when lung transplantation becomes contraindicated (8).
- Do-not-resuscitate (DNR) orders are often not written until the final few days of life. Many patients continue to receive disease-modifying therapies up until the last 24 hours of life (9). Although many disease-specific therapies can be administered in the home setting, most CF patients die in the hospital setting (5). This likely reflects a combination of prognostication challenges, reluctance to abandon aggressive therapies, patient/family preference, and caregiving burdens. CF patients may be uniquely suited to Medicaid ‘concurrent care’ waiver programs in which patients can receive hospice and routine medical care simultaneously (10).
- Although guidelines are lacking, progressive hypoxic respiratory failure may benefit from long-term oxygen therapy (6). Non-invasive positive pressure ventilation (NIPPV) has a role for hypercapneic

respiratory failure, dyspnea management, and, in select candidates, as a bridge to lung transplant. Use of invasive positive pressure ventilation (IPPV) should always involve clear discussions around therapeutic options and goals-of-care (8).

- In the past, intensive care for CF patients was generally considered futile. More recently, ICU outcomes have improved, particularly for patients requiring NIPPV (as opposed to IPPV), and for patients who are candidates for lung transplantation (6,8). Long-term outcomes for respiratory failure requiring IPPV remain dismal however, and intensive care management may commit many patients to eventual end-of-life care in the ICU setting.
- Common symptoms at the end of life include dyspnea, fatigue, anxiety, anorexia, pain, and cough (see *Fast Facts* #27, 199, 200). Care providers must balance benefit vs. burden of disease-specific treatments such as nebulized medications, NIPPV, and chest physiotherapy (6,8).
- Patients who survive multiple episodes of acute respiratory failure may come to overestimate their resilience, making appropriate goals-of-care discussions or end-of-life care planning more challenging (6,7).

Key teaching points

- Pulmonary involvement in CF is the most common cause of morbidity and mortality in people affected by this disease. Symptoms such as dyspnea, anxiety, cough, and pain commonly interfere with CF patients' quality-of-life. For some patients, lung transplantation offers hope of improved survival and quality-of-life. However, most will die of their disease in the hospital setting.
- Accurate temporal prognostication in CF is challenging. However, the natural history of CF without transplantation is well-established and patients and families should be prepared for the physical and emotional challenges of end-stage CF, including end-of-life decision-making, even without a precise estimation of survival time.

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