Journal of the American Pharmacists Association 58 (2018) 246–249

Contents lists available at ScienceDirect

Journal of the American Pharmacists Association

journal homepage: www.japha.org

COMMENTARY

The pharmacist's role in supporting people living with cystic fibrosis

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ARTICLE INFO

Article history:
Received 24 August 2017
Accepted 20 January 2018

ABSTRACT

Objectives: To describe the critical need for pharmacists' involvement in outpatient care for people living with cystic fibrosis (CF).
Data sources: Not applicable.
Summary: CF is a pulmonary condition that affects more than 30,000 children and adults in the United States and 70,000 people worldwide. Various complex medication regimens are given to patients with CF, some depending on the type of mutation they have in their CF transmembrane conductance regulator protein. With complex medication regimens and the increased number and variety of treatments that have become available, the medication use burden intensifies for individuals living with CF and their caregivers. Young people living with CF have a particularly difficult time adhering to medications and other therapies as they begin to rely less on their caregivers and assume greater medication management responsibility for their care. Adolescents report low adherence rates from about 40% to 47% for airway clearance methods and even lower for nutritional recommendations, about 16% to 20%. In inpatient settings, pharmacists have been successful in making medication use recommendations that have improved adherence for patients with CF while in the hospital. However, limited research has explored how provision of pharmacist supportive care and patient education in outpatient settings can improve medication adherence and quality of life for people living with CF.
Conclusion: There is potential for provision of outpatient pharmacy clinical services to increase medication adherence and overall quality of care for patients with CF. Higher rates of medication adherence in patients with CF could in turn improve patient outcomes and reduce overall health care costs as a result of fewer rehospitalizations. Pharmacies can implement programs designed to provide comprehensive support services and medication management from pharmacists and staff that are trained in CF care.

Cystic fibrosis (CF) is one of the most common complex, life-shortening, and fatal inherited diseases and presents a considerable management challenge to health care providers.[1,2] This chronic genetic condition primarily affects the respiratory and digestive systems.[3] CF is a progressive multisystem disorder characterized by abnormalities in the transport of chloride ions in human airway epithelial cells, resulting in an ionic imbalance that impairs the clearance of secretions, or mucus, primarily in the lungs, small intestine, liver, and pancreas.[4] Thick mucus production can lead to frequent lung infections, decreased pulmonary function, inability to properly digest food and absorb essential nutrients, and complications with other organs.[4] Lung transplantation remains an option for some of these patients, because they often experience progressive lung damage, which results in increased rates of morbidity and mortality. More than 30,000 children and adults in the United States have CF (more than 70,000 worldwide), and the average life expectancy for people living with CF who were born from 2012 to 2016 is 43 years.[5] This is a 12-year increase in life expectancy compared with the average life expectancy of 31 years for people living with CF who were born from 1992 to 1996.[5] Treatment advances

Disclosure: The authors declare no relevant conflicts of interest or financial relationships.

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https://doi.org/10.1016/j.japh.2018.01.006

Key Points

Background:

- Cystic fibrosis (CF) is a complex genetic disease that presents a considerable management challenge to health care providers, caregivers, and patients.
- On average, medication regimens for patients with CF include about 8 to 10 medications daily. The medication regimen is complex and can take 1 to 3 hours to complete.
- Consequently, patients with CF encounter unique and significant challenges with managing their daily medication regimens, which contributes to high rates of nonadherence.

Findings:

- Pharmacists can support patients with CF by working with them to tailor their medication regimens to meet their individual needs, because of their expertise in pharmacotherapy, pharmacodynamics, and drug interactions.
- There is a need for patient education and supportive services for people living with CF within outpatient pharmacy settings to improve daily medication adherence, reduce the number of hospitalizations, enhance their quality of life, and extend their life span.

paired with aggressive management have increased life expectancy in the past 30 years.[6]

Management of this chronic condition requires complex lifelong medication regimens that pose significant challenges for patients with CF and their informal caregivers.[1,7] Advances in the knowledge and treatment of CF have led to more people being diagnosed with CF during infancy and living into adulthood.[4] There is currently no cure for CF; therefore, those living with this chronic condition are faced with burdensome medication management requirements that can take 1 to 3 hours each day.[7,8] Most patients with CF and their caregivers report that it is not feasible to travel long distances for routine outpatient care; therefore, there is a need for more supportive care that is easily accessible to their homes.[9] In addition, patients may have monthly copayments ranging from $107 to $310 for the prescription medications they take each day.[10] These combined challenges contribute to poor medication adherence, which results in increased health care costs, exacerbations, depression, anxiety, and hospitalizations.[8,11-13]

Patients with CF often face many psychosocial challenges. Owing to their increased risk of infection and cross-contamination, many patients are isolated from others and frequently report high levels of depression and anxiety, which greatly affect disease severity and outcomes, treatment adherence, self-management, and understanding of CF.[2,14] Ideally, people living with CF should receive multidisciplinary specialized inpatient and outpatient care from CF Foundation (CFF)—accredited centers. Dedicated CFF-accredited centers where patients receive specialist care are associated with improved survival and quality of life.[15,16] Patients receive frequent clinical evaluations, monitoring, and education from these care center visits where all of the health care workers are specifically trained in CF management and treatment.[17] Standards of care have been defined for these centers to ensure the best outcomes possible for patients.[17] CFF-accredited centers are required to have the staff and facilities to provide care and treatment for all CF-associated complications.[17] According to American CF standards of care, CFF-accredited centers should require many CF specialists, including nurses, dietitians, physicians, social workers, respiratory therapists, and program coordinators.[18] Pharmacists are included as recommended team members for CFF-accredited centers.[18]

There is increasing awareness regarding the necessity for pharmacists to care for CF patients, not only in the inpatient setting, but particularly in outpatient settings. Consequently, the CFF recently awarded funding to more than 50 CFF-accredited centers to increase pharmacy services in the outpatient setting.[19] To our knowledge, very little is known about the role of pharmacists in outpatient settings for people living with CF. Within inpatient settings, the CF clinical pharmacist educates patients and caregivers about proper administration of inhaled medications, alternate or less expensive therapy that may be available, and the best order and timing for patients to take their medications.[17] In addition, the pharmacist obtains and documents accurate medication lists for patients, focusing on access and adherence, and recommends changes according to CFF guidelines. The pharmacist also monitors for drug interactions, adverse effects, and drug-nutrient interactions in patients' complex medication regimens, which include many inhaled, intravenous, and oral medications.[17] The pharmacist is particularly useful for the proper dosing of antibiotics, such as aminoglycosides, to ensure minimal toxicity and maximum therapeutic benefit for each patient.[17]

Complexity of CF medication use

Treatment burden and subsequent nonadherence are key issues that affect daily CF management.[8] Medication use challenges and errors are a serious patient safety problem for patients with CF because they use about 8 to 10 medications daily.[20,21] Current CF therapies include CF transmembrane conductance regulator protein (CFTR) modulators, inhaled antibiotics, pancreatic enzymes, supplemental vitamins, supplemental calories, inhaled mucolytics, antiinflammatories, and pulmonary physiotherapy.[22] The modulator therapy works to correct specific mutations among the 5 different classes of CFTR mutations.[22] Accumulation of mucus in the lungs increases CF patients' susceptibility to bacterial lung infections, so many patients take inhaled antibiotics as cycled therapy.[22] Pancreatic enzymes are taken with all food and supplemental intake to promote digestion and nutrient absorption, and specific diet changes are recommended.[22] Inhaled mucolytics are aerosolized agents that thin the mucus in the lung airways to help with mucus removal when coughing.[22] Antiinflammatories are used to reduce the excess mucus in the lung airways.[22] Treatment complexity presents numerous barriers to optimal treatment, including low motivation and poor adherence,[8,14] adverse effects, suboptimal dose delivery