Original Article

Descriptions of the Pain Experience in Adults and Adolescents with Cystic Fibrosis

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Abstract:

People living with cystic fibrosis experience pain that is associated with decreased quality of life, poorer health outcomes, and increased mortality. Though pain is highly prevalent as a symptom, it is currently unknown how persons with CF describe their pain experiences or the ways those experiences impact their lives. To explore and describe ways adolescents and adults with CF experience pain. An exploratory descriptive design was implemented to perform interviews with 10 individuals with CF and self-reported moderate to severe pain. The interviews explored their pain experiences within five domains: Pain Characteristics, Activities, Relationships, Work/School Life, and Health Care Team. Transcribed interviews underwent a content analysis with team-based constant comparisons. Individuals with CF identify the disease as being painful; express how pain negatively affects all aspects of their lives, including loss of functionality and productivity; and are able to disclose their pain to those with whom they have relationships. Adolescents feel an emotional toll from the loss of socialization as a result of pain and feel their health care team adequately supports their pain. Adults express a unique emotional pain component to CF and feel stigmatized and unsupported by their health care team when asking for pain management solutions.There are differences in how pain is perceived by adolescents and adults with CF that have otherwise not been reported in the current literature. Further explorations of pain across the lifespan and health care provider attitudes toward pain management are needed to guide the development of effective pain management interventions for those with CF.

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Pain is a complex, multidimensional process that negatively affects physical and mental functioning, clinical outcomes, quality of life, and productivity (Schiller, Lucas, Ward, & Peregoy, 2012). The prevalence of pain in the general population

is widespread and can account for up to 80% of all general practitioner visits (Jamison & Edwards, 2012). The effective treatment of chronic pain—defined as persistent unpleasant sensory and emotional experience lasting more than 6 months and without an anticipated or predictable end (International Association for the Study of Pain [IASP], 2014)—is a public health concern associated with loss of productivity, loss of functionality, increased risk for mental health conditions, with significant costs to the health care system (Stockbridge, Suzuki, & Pagan, 2015).

Cystic fibrosis (CF) is a life-limiting autosomal recessive disease affecting approximately 30,000 people in the United States and 60,000 people worldwide (Cystic Fibrosis Foundation [CFF], 2016). The disease is characterized by recurrent respiratory infections, pancreatic insufficiency, CF-related diabetes, sinus disease, and other complications such as absence of the vans deferens in males (Davis, 2006). No longer considered a terminal childhood disease, advances in treatments and a greater understanding of the underlying genetic mutations responsible for CF have increased the life expectancy from an average age of 6 months in 1938 (Davis, 2006) to the current average age of 43 years (CFF, 2016). Despite these advances, individuals with CF still experience considerable physical (e.g., pain, cough, dyspnea, fatigue) (Goss, Edwards, Ramsey, Aitken, & Patrick, 2009) and psychological symptoms (e.g., depression, and anxiety) (Quittner et al., 2014) that are associated with increased disease severity and decreased quality of life (Festini, Ballarin, Codamo, Doro, & Loganes, 2004; Havermans, Colaert, DeBoeck, Dupont, & Abbott, 2013; Quittner et al., 2014; Sawicki, Sellers, & Robinson, 2008).

Symptom management in those with CF largely focuses on the treatment of the underlying pathologic condition as opposed to direct symptom-managing interventions. In one study, 94% of surveyed individuals with CF reported a pain event within a 2-month period, with one third of those reporting it as moderate to severe (Festini et al., 2004). However, there are no standard assessments, measures, or treatments for pain in this population (Havermans et al., 2013). Although multiple retrospective studies have reported on the location, duration, and severity of pain in both adolescents and adults with CF and its negative relationship to health-related quality of life and disease outcomes (Blackwell & Quittner, 2014; Hayes et al., 2011; Hubbard, Broome, & Amita, 2005; Koh, Harrison, Palermo, Turner, & McGraw, 2005; Lechtzin et al., 2016; Palermo, Harrison, & Koh, 2006; Perquin et al., 2000; Kelemen et al., 2011; Sermet-Gaudelus et al., 2009), the role of pain in the disease process, its effect on daily life, and how the impact of pain changes across the lifespan are poorly understood. Understanding patient pain characterization and the consequential limitations the pain experience places on individuals with CF is essential to developing a standard, clinical approach to CF pain management and treatment.

The purpose of this study was to explore patient-reported descriptions of the pain experience among adolescents and adults living with CF.

METHODS

Design

An exploratory descriptive design was used to gain firsthand knowledge of individual experiences with pain (Sandelowski, 2009).

Setting and Sample

Study participants were recruited from a nationally accredited Cystic Fibrosis Care Center within a large teaching hospital's outpatient specialty clinics. The center is composed of a pediatric clinic and an adult clinic that sees approximately 550 patients with CF per year. Eligible participants were identified through a positive response to the presence of moderate to severe pain occurring at least once a week for a period of at least 1 month on the Brief Pain Inventory during previous participation in a self-reported pain survey.

A purposive sample of 10 participants were enrolled into the study. Other inclusion criteria were that the participants had a diagnosis of CF and could understand English in order to provide informed consent. Participants who had undergone a solid organ transplant were excluded from the study. All participants who were invited agreed to participate. Participants received a parking voucher as a reimbursement for their participation.

Procedures

The Johns Hopkins Medicine Institutional Review Board (IRB00033661/NA_00027981) approved the study. Written informed consent was obtained from adult participants, and written parental consent with adolescent assent was obtained from adolescent participants per the institutional review board requirements. Informed consent was obtained at outpatient clinic visits, and individual in-depth telephone or face-to-face interviews were scheduled for each participant, depending on participant availability and clinic space. Demographic data and participant characteristics were collected from electronic medical records.

A semistructured interview guide was developed by the study team through formative exploration of the available literature, clinical experiences, and