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

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Abstract

Background: 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.

Aims: To explore and describe ways adolescents and adults with CF experience pain.

Design/Setting/Subjects/Methods: 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.

Results: 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.

Conclusion: 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.

Pain is a complex, multidimensional process that negatively affects physical and mental functioning, clinical outcomes, quality of life, and productivity (Schiller, Lucas, Ward, &

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DISCLOSURES

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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 vas 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 informal discussions with providers and patients. The characterization of pain and the effect of pain on quality of life, productivity, and relationships were identified as gaps in the current CF literature that could be addressed by this study. Additionally, patients with CF and nurses who specialize in CF care expressed the need to explore how pain is assessed, addressed, and treated by health care providers. Both adolescents and adults received the same questions, with school life being inserted in place of work life where applicable. The domains and questions are displayed in Table 1.

Audio-recorded telephone or face-to-face interviews were conducted by two different interviewers relative to their adolescent and adult group-specific experience. Data collection occurred over an 8-month period. Each audio-recorded interview was transcribed verbatim and crosschecked for accuracy, and then transcripts were deidentified before analysis.

Data Analysis

The collected data were analyzed via a conventional content analysis (Hsieh & Shannon, 2005) and managed using the NVivo qualitative research software package, Version 10.2.1. Transcripts were first read independently by each team member several times to allow for general impressions of the content to develop into categories and preliminary codes. The team then met to discuss the independently coded content. Discrepancies were reconciled through team consensus for each emergent code. Previously coded transcripts were compared with each newly coded transcript to ensure consistency in code assessment (Sandelowski, 2000). Peer review of the analysis was performed by clinical and research experts in CF at conclusion to ensure trustworthiness (Creswell, 2007; Polit & Beck, 2010).

RESULTS

Participant demographic data including age, sex, and disease severity—represented as forced expiratory volume in 1 second percent predicted (FEV1%)—and accompanying comorbidities commonly associated with CF were collected (Table 2). Of the 10 participants enrolled, one participant was withdrawn because the person underwent a lung transplant between the time of enrollment and the scheduled interview. The age of participants ranged from 13–46 years with 50% of the participants being female. FEV1%, a value typically indicative of obstructive pulmonary disease severity, ranged from 23.5% to 102.0%, with the adult FEV1% trending more severe compared with the adolescent FEV1% range. Pain location was reported by participants as the lungs, head, sinuses, abdomen, back, ribcage joints, and whole body.

Concordant and discordant themes between the adolescents and adults emerged from within the explored domains. The adolescents discussed the effects of pain on their social lives and overall felt supported by their health care team in terms of pain management. The adults discussed the pervasive nature of their pain, the emotional pain associated with CF, and the barriers within the health care system that contribute to poor pain management. All participant quotations have been ascribed to a pseudonym.

Recognizing CF-Associated Pain

The ability to differentiate between CF-specific pain and other, more typical sources of pain—for example, a strained muscle after a strenuous workout—emerged as a strong concordant theme between the two groups. The participants clearly distinguished the pain they associate with their CF as a separate and unique pain:

“I also have kind of like CF-related pain too.” (Don, 13 years)

“...Yeah, I think it’s...definitely a different pain.” (Roy, 32 years)

When asked about the quality and locations of the pain they experience, adolescent participants reported experiencing pain in episodes and were not able to verbalize pain duration but were able to attribute their pain more to a specific event:

“It’s usually in my stomach because I forget my enzymes.” (Josh, 15 years)

“You know my back pain they have said that it’s just from coughing.” (Susan, 19 years)

“Aching,” “shooting,” “stabbing,” and “constricting” were the terms used to describe the quality of their pain.

Adult participants reported experiencing pain that is pervasive in duration:

“I would say 3 or 4 years now.” (Tom, 30 years)

It was more difficult for the adult participants to find single descriptive words for the quality of their pain compared with the adolescent group:

“It’s just a consistent, almost like tightness, umm, it—at any given point it’s not overwhelming or stabbing or sharp or anything like that, it’s just a consistent, um, I always have trouble describing it—like a tightness, um, sometimes it feels almost like somebody’s reaching up under my ribcage and pulling outwards.” (Paul, 34 years)

A subtheme, *Recognizing Emotional Pain*, emerged within the adult group. Adult participants reported they experience more than physical pain and discussed how depression and emotional pain is a significant aspect of their CF disease:

“Emotionally...is equal to the physical pain, and often is much more harming or hurtful.” (Molly, 46 years)

“It’s an emotional rollercoaster.” (Joan, 34 years)

“It’s a big part of CF is emotional pain, and depression.” (Roy, 32 years)

Restrictions of Pain on Life

The negative impact of how pain restricts daily living, recreational, and work-related aspects of life was shared among all participants. Participants described how pain limits their ability to function and perform typical activities of daily living, revealing that on some days their pain is so severe it prevents them from getting out of bed:

“Once I wake up, the first hour I just sit on the couch...just wait until I feel better.” (Tim, 30 years)

“Especially some days because the pain gets really bad it just—I just on really bad days I can’t get out of bed.” (Terry, 16 years)

“I don’t feel well enough to get up and move around. Sometimes I can’t even get out of bed.” (Molly, 46 years)

Participants also shared how pain restricts their ability to participate in social and family recreational activities. Family vacations, hobbies, and planned social events have to be modified or cancelled because of the limiting effects pain has on their ability to participate and enjoy in these activities:

“I left my sophomore homecoming because I didn’t feel good...being on my feet and stuff like that was like really hurting me. So my mom, um, my mom came and picked me up from the dance and I just didn’t really do anything, I just came home

and my friends went, out afterwards and hung out together and whatever...”
(Monica, 19 years)

Additionally, participants described how pain affects their ability to keep up with their workload from a functional perspective, but also from the effects of absenteeism:

“I usually have tests like every other day...and I remember, ‘Oh wait, I’m going to have to catch up on everything’ and I’m like Blergh!” (Donny, 13 years)

“I usually just work like 2 days a week, 6 hours each day. That’s just because anything else will wear me out really.” (Paul, 34 years)

A subtheme, *Social Life and Emotional Toll*, emerged from the adolescents we interviewed. There is an emotional toll from missed sporting events, social activities, and school functions that is otherwise not shared by the adults we interviewed:

“I was pretty bummed out.” (Josh, 15 years)

“...I try and understand it, but it’s kind of upsetting sometimes.” (Sue, 14 years)

Sharing the Pain

The willingness to disclose pain to family, friends, and people at work/school was shared by the participants. They reported that families and close friends are supportive and understanding of how pain affects and limits their ability to function in day to day life:

“I can pretty much talk to him about anything, whether it’s pain, or CF-related...”
(Molly, 46)

“Most of my friends, like, they understand.” (Donny, 13)

“For a while I was like, No, I don’t need anybody, I don’t need anybody to talk to, I don’t need anybody to talk about my pain and now, it’s like I want to tell everybody!” (Monica, 19)

All participants discussed that they disclose their pain to bosses and teachers, though one adult says he volunteers the information with hesitation mostly due to internal fear of job loss:

“You’re scared that you’ll lose your job...in general no one has ever made me feel like I have to hide it. It comes from me mostly.” (Roy, 32)

Feeling Supported and Treated.—All participants spoke to the fact that they do discuss their pain with their CF health care providers, but there were stark differences in the perceptions of how pain is addressed and treated between the groups. Adolescent participants had a strong belief that their health care providers were open to hearing about their pain and were helpful in finding treatment options, and they expressed feeling supported and satisfied with the relationship:

“They always want to know my side of the story...they know how to handle [it] in my view at least.” (Terry, 16)

“What they’re doing is pretty good.” (Josh, 15)

Feeling Disbelieved and Stigmatized.—The adult participants expressed feeling that the health care providers who treated them CF did not always believe in their CF-specific pain, they did not feel that their pain needs were being met, and they felt stigmatized when asking for pain medications:

“They weren’t 100% sure, um, that what I was describing was, you know, actual physical pain, um, it took a lot, took a little bit for them to come around and realize it.” (Tim, 30)

“But it kind of made me feel really bad about myself...I was like a wimp and I should be sucking it up and handling it because I was a CF patient, and we’re not supposed to have pain...you feel like going to the doctors asking for pain meds, you feel like you’re like a druggie ’cause you’re like, ‘No that doesn’t work anymore.’” (Molly, 46)

DISCUSSION

The participants were very forthcoming in their discussions around pain, and many expressed thanks and relief in having a voice for their pain and how it affects their lives. They provided rich insights into their pain experiences, illustrating how pain is burdensome on emotional, social, and functional levels. In particular, the ability of individuals with CF to differentiate “normal pain” from CF-specific pain has not before been reported. Some of the descriptive phrases the participants used when discussing their physical pain, including “stabbing,” “constricting,” and “pulling outwards,” provide vivid details about the experience of pain that is otherwise not captured by quantitative studies. CF is a multisystem disease that can be responsible for painful inflammatory and endocrine responses such as bone loss in osteoporosis or pancreatitis as a result of pancreatic insufficiency (Davis, 2006). The pain experienced by those with CF is not limited to one specific body system, which suggests that multiple mechanisms of action are responsible for their pain.

Studies by Hayes et al. (2011) and Lechtzin et al. (2016) have associated pain with worse clinical outcomes in patients with CF, including more hospitalizations and increased risk for mortality. This is not surprising given the bidirectional relationship between pain and immune function (Woda, Picard, & Dutheil, 2016). These findings indicate that patients perceive a pain that is uniquely specific to their CF disease process, suggesting that pain should be assessed, addressed, and treated not as an independent entity but as a symptom or comorbidity of CF.

The negative association of pain and quality of life has been well documented in cohort studies of patients with CF (Havermans et al. 2013), and both adolescent and adult participants in this study endorsed the disabling effects of pain on function, including performing activities of daily living. What has not been previously documented, however, is the debilitating influence pain has on all aspects of their lives. Our study revealed specific ways and areas in which individuals with CF are restricted by their pain. Participants described how work and school became challenging, if not impossible; recreational activities were put on hold or avoided; and when the pain was at its worst, they were bedbound and unable to ambulate. Despite the *Feeling Supported and Treated* theme that emerged among

the adolescent participants, the loss of functional ability that was revealed suggests that this may not be the case. There are currently no standards of care specifically for pain management in this population. Episodic pain, such as abdominal pain as a result of nonadherence to pancreatic enzymes or a headache caused by sinusitis, can be well managed through treatment of the underlying cause. Hot packs and acetaminophen are routinely ordered when patients are hospitalized, but the amount and depth of education received for self-management of pain at home is unknown. Pervasive pain, as discussed by the adults, is more difficult to manage. The experiences of participants feeling stigmatized by seeking pain relief, health care provider disbelief of the presence of physical pain, and uncertainty in how pain should be addressed in patients with CF indicates a lack of knowledge of pain management options within the health care system. These findings warrant immediate attention to increasing pain management competencies within health care providers who specialize in CF care and to exploring the use of integrated symptom management specialists into CF standards of care.

The emergence of the *Social Life and Emotional Toll* subtheme from the adolescent participants is consistent with the developmental stage of this age group (Fogeron et al, 2010). Peer groups and social events are the focus of an adolescent's life. The episodic and unpredictable nature of their pain (Blackwell & Quittner, 2014; Lechtzin et al., 2016; Palermo et al. 2006; Sermet-Gaudelus et al, 2009) may place these participants at a greater risk for compromised social functioning. Provider and patient education regarding strategies for self-management is needed to help alleviate pain as it is occurring and before it becomes a hindrance to activities.

Both adolescents and adults expressed an openness and willingness to disclose pain to those in most aspects of their lives, including their families, friends, peers, and professional colleagues. A 2010 study by Modi et al. found that 94% of adults with CF are comfortable disclosing their CF status to relatives, 81% to close friends, and 51% to bosses and teachers (Modi, Quittner, & Boyle, 2010). The theme of *Sharing the Pain* In our study is consistent with these findings given that the participants we interviewed consider pain to be a part of their CF disease.

The subtheme of *Recognizing Emotional Pain* that emerged from the adult interviews is not surprising given recent data on the prevalence of anxiety and depression among persons diagnosed with CF (Quittner et al., 2016; Screening & Treating Depression & Anxiety Guidelines, 2015) and the known association between pain and depression/anxiety (Hayes et al., 2011). The discussion of emotional pain also suggests that there is a psychosocial health need in the adult population that is not being met by current standards of care. The CF Foundation recently released clinical care guidelines recommending the use of depression and anxiety screening tools as a standard of care for patients with CF (Quittner et al., 2016) in an effort to help identify those who may be experiencing emotional distress or have a mental health disease. Qualitative studies exploring the sources and perceptions of emotional pain should be undertaken to guide the implementation of clinical interventions.

The adult participants also noted that they did not feel supported in their attempts to discuss their pain, nor did they receive treatment or adequate treatment from their health care

provider. These findings bring to light the general stigma that exists in the health care community around pain treatments and drug-seeking behavior (Meltzer et al., 2013) as well as the importance of implementing multidisciplinary teams in the treatment of life-limiting chronic diseases. Future qualitative studies of provider beliefs, management practices, and understanding of pain are needed. This information could be used to guide the development of educational programs geared toward those who provide care to individuals with CF.

Limitations

The sample size was limited to 10 patients with moderate to severe self-reported pain. Though the prevalence of pain associated with CF can be as high as 94% of the population (Festini et al., 2004), the intensity and duration is often much less than what is represented by this purposively sampled group. Additionally, there is a paucity of current evidence on this topic, as reflected by the extended use of citations more than 5 years old. Research involving pain in individuals with CF has been conducted sparingly over the years and at single care centers. Larger studies exploring the development of pain across the lifespan, investigating the current use and effectiveness of pain interventions, and developing an understanding of the CF-specific disease factors that could be related to pain development are needed in this population.

CONCLUSION

This study provided much needed insight into the pain experiences of adolescents and adults with CF. CF pain is perceived to be different from other types of pain, the impact on all aspects of life is severe enough to inhibit activities of daily living, and individuals with CF are willing to talk to others about their pain. Adolescents experience episodic pain, feel supported in their pain management, and are at risk for impaired social functioning as a result of the emotional impact of missed social events. Adults experience persistent pain, recognize a distinct emotional pain associated with CF, and feel stigmatized and unsupported in their attempts to seek pain treatments. A larger qualitative study of pain in persons with CF as well as studies exploring the beliefs, knowledge, and competencies of pain management in health care providers are warranted to provide much needed insight into the impact of pain during key developmental stages and to help guide the development of education and treatment interventions for pain management in this population.

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Table 1.**Interview Questions**

Question Domain	Questions
Pain Characteristics	Are you currently having any pain? Usually, where is your pain located? How would you describe the pain (for example, stabbing, dull, throbbing etc.)? How long have you been experiencing this particular pain? Do you currently take any medications or undergo any therapies to help alleviate your pain? If yes, please explain and rate effectiveness, if drug/alcohol use is endorsed by participants freely then follow up questions regarding effectiveness may be pursued,
Activities	Can you tell me a little bit about your home life (living arrangements, family, etc.)? What kinds of things do you like doing for fun? How does pain affect your ability to do those things? Tell me about a time that pain has prevented you from doing something you wanted to do. How does pain, if at all, limit your ability to perform your activities of daily living (brushing teeth, CF treatments, eating, etc.).
Relationships	Tell me about your friends/family and your comfort discussing your illness with them. Describe how you talk about your pain with your family/friends Tell me about a time your family/friends responded (can be negatively or positive through support) to a situation where your pain limited you from doing something with them
Work/School Life	Describe your work/school life (if currently not working, explore if illness/pain is the reason why and discuss previous work experiences and how CF and pain affected performance) How comfortable are you discussing your illness and pain with people at work? Tell me about a time pain affected your ability to work and how your work responded. Describe to me your level of comfort in telling your work about your CF and your pain
Health Care Team	How comfortable are you discussing your pain with your healthcare team? Describe to me how your pain is addressed/treated in clinic Have you had any experience as an inpatient? If yes, describe your experiences with the inpatient team and how your pain is addressed

CF = cystic fibrosis.

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Table 2.

Baseline Characteristics

	Adolescents, n = 5	Adults, n = 5
Age (range)	13–19 years	30–46 years
Sex (female)	3 (60%)	2 (40%)
FEV1 (% predicted range)	53.0%–102.0%	23.596–82.3%
Pancreatic insufficient (%)	5 (100%)	5 (100%)
Cystic fibrosis–related diabetes (%)	2 (40%)	1 (20%)
Osteopenia/osteoporosis (%)	1 (20%)	4 (80%)
Sinus disease (%)	2 (40%)	3(60%)
Depression (%)	0 (0%)	2 (40%)
Anxiety (%)	0 (0%)	1 (20%)

FEV1 = forced expiratory volume in 1 second percent predicted.