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The Influence of Pain Catastrophizing and Parent Behavior on Health-Related Outcomes for Youth with Sickle Cell Disease

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by

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ABSTRACT

Youth with sickle cell disease (SCD) experience frequent pain, which is related to their functioning and health-related quality of life (HRQoL). A growing body of literature suggests that rather than the experience of pain experience itself, the cognitive appraisal of pain is critical. The overall aim of the current study was to examine the relationship between child pain catastrophizing, parent response to child pain symptoms, and health-related outcomes (i.e., functional disability, health-related quality of life; HRQoL). Results indicated that pain catastrophizing significantly predicted functional disability and HRQoL above and beyond children's experience of pain. Additionally, protective, minimizing, and encouragement/monitoring parent responses each moderated the relationship between pain catastrophizing and HRQoL; for youth who engaged in moderate to high levels of pain catastrophizing, parent response to pain appeared to be more impactful on HRQoL. Clinical implications regarding intervention for pain catastrophizing and parent response to pain are discussed.

INDEX WORDS: Pediatric, Sickle cell disease, Pain, Pain catastrophizing, Parenting, Health-related quality of life

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DEDICATION

To my mother, my father, and my sister Elizabeth, this project would not have been possible without your unconditional love and support. Thank you for inspiring me, believing in me, and providing endless encouragement. Thank you for your sacrifices that have allowed me to achieve.

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1 INTRODUCTION

1.1 Overview of Pediatric Sickle Cell Disease

Sickle cell disease (SCD) is one of the most commonly inherited blood disorders in the United States, affecting about 100,000 individuals (Centers for Disease Control and Prevention, 2020), and global prevalence is expected to increase in the next thirty years (Kapoor et al., 2018), due to increased survival rates of infants born with SCD and increased life length of individuals with SCD. In children under the age of five, it is estimated that the global prevalence of sickle cell anemia (SCA), the most severe form of SCD, will increase by thirty percent in the next forty years (Piel et al., 2013). Individuals born with SCD have red blood cells that take on an abnormal or "sickled" shape, which affects the cells' ability to carry and distribute oxygen (National Heart, Lung, and Blood Institute, 2018). This sickle shape also causes red blood cells to adhere to each other, which ultimately can lead to the obstruction of blood vessels throughout the body, thereby decreasing or eliminating blood flow to vital organs. Therefore, patients with SCD are at elevated risk for medical complications from disease pathophysiology, which can lead to infections, acute and chronic pulmonary complications, bone infarctions, organ failure, and stroke (Ballas, 2007; National Heart, Lung, and Blood Institute, 2018; Platt, 1991). In addition, there can be complications associated with disease management, which can include risk for transfusional iron overload, alloimmunization, transmission of infections, and other issues (Kapoor et al., 2018).

Individuals who are carriers of a single sickle cell gene are referred to as having the sickle cell trait (SCT) and appear to have some protection against severe malaria infection (Platt et al., 2011). Thus, historically, SCD has been geographically concentrated in tropic and sub-

tropic areas of the world, where malaria was endemic. SCD is most commonly found in individuals of African, South or Central American, or Mediterranean descents. In the United States, SCD disproportionately affects the Black or African American community, occurring in 1 of 365 births (National Heart, Lung, and Blood Institute, 2018).

The most common and challenging symptom of SCD is recurrent and unpredictable vaso-occlusive pain episodes (Centers for Disease Control and Prevention, 2020). Pain, defined as "an unpleasant sensory and emotional experience associated with, or resembling that associated with, actual or potential tissue damage," is inherently influenced by biological, psychological, and social factors (International Association for the Study of Pain, 2020). Therefore, an individual's experience of pain is largely determined by individual factors, thus subjective in nature and challenging to quantify. Pain frequency and severity varies significantly across and within individuals. Although most pain episodes are mild to moderate and may be managed at home, severe pain often warrants hospitalization. Largely, ongoing clinical research efforts target advancements in SCD pain and symptom management. These efforts have allowed for diversity in disease management and treatment options, which has increased survivorship, warranting greater research and clinical efforts to bolster quality of life and functional outcomes.

1.2 Health-Related Outcomes in Pediatric Sickle Cell Disease

Respective to other pediatric chronic illness populations, there is a limited body of research that has examined health-related outcomes, specifically functional and psychosocial outcomes, for youth with SCD. However, it is broadly understood that youth with SCD are at increased risk for worse outcomes when compared to healthy peers (e.g., Anie, 2005; Sil et al., 2016). Given pain, hospitalizations, and other symptoms, youth with SCD report mild to moderate impairment in their abilities to engage in daily activities at home, at school, in recreational activities, and during social interactions, which is commonly described as functional

disability (Oliver-Carpenter et al., 2011; Palermo et al., 2008; Peterson & Palermo, 2004; Sil et al., 2016; Walker & Greene, 1991). Preliminary investigations in this population suggest that the frequency of youths' pain is associated with severity of functional disability, such that youth who experience pain three or more days each week experience greater levels of impairment, respective to youth who experience pain less frequently (Sil et al., 2016). When compared to other pediatric chronic illness populations, youth with SCD report rates of functional disability greater than youth with other diseases, including juvenile idiopathic arthritis, but did not report as severe impairment as youth with chronic headaches (Peterson & Palermo, 2004). In a cross-sectional investigation of youth with SCD, physical limitations (e.g., running, walking, playing sports) were rated as most challenging for youth (Oliver-Carpenter et al., 2011).

With respect to psychosocial functioning, the extant literature is mixed with regard to whether youth with SCD report elevated rates of depressive symptoms. Some investigations have documented greater rates of depressive symptoms and disorders in youth with SCD when compared to healthy peers (Jerrell et al., 2010; Morgan & Jackson, 1986; Yang et al., 1994), whereas others have observed no significant difference (Graves et al., 2016; Kumar et al., 1976; Lee et al., 1997; Sil et al., 2016; Simon et al., 2009). In the adult SCD literature, investigations have consistently reported that adults with SCD experience more depressive symptoms than their non-SCD counterparts (Jonassaint et al., 2016; Laurence et al., 2006; Molock & Belgrave, 1994). Though these studies are cross-sectional in nature, it is hypothesized that unpredictable, recurrent pain may contribute to feelings of hopelessness and lack of control in adults, which may also provide insight into youths' pain-related cognitions and how they impact psychosocial functioning (Molock & Belgrave, 1994).

Data also indicate that youth with SCD perceive the effect of their chronic illness on their physical, mental, and social aspects of their well-being, also referred to health-related quality of life (HRQoL; Karimi & Brazier, 2016) as poor, especially when compared to healthy peers (Dale et al., 2011; Palermo et al., 2002; Panepinto et al., 2005). In a study comparing HRQoL across pediatric chronic illness populations, youth with SCD had one of the lowest scores, which suggests that youth with SCD perceive their disease to impact their quality of life more negatively than youth with many other chronic illnesses including cystic fibrosis, irritable bowel disease, epilepsy, type 1 diabetes, kidney transplant recipients (Ingerski et al., 2010). Studies suggest that frequency of pain episodes, disease severity, and other SCD-complications are associated with lower HRQoL (Dampier et al., 2010; Fisak et al., 2012; Panepinto et al., 2005; Panepinto et al., 2009). Palermo et al. (2002) suggest that parents of children and adolescents with SCD reported low HRQoL, which is associated with poor self-esteem, school participation, social engagement, and psychological adjustment.

1.3 Pain, Pain Catastrophizing, and Health-Related Outcomes

Across illness populations that experience pain, the literature suggests that the frequency and intensity of pain is associated with worse health-related outcomes (Bakshi et al., 2018; Barakat et al., 2008; Jackson et al., 2014). Specifically, it is documented that youth who experience more frequent pain (Brandow et al., 2009; Huguet & Miro, 2008; Sil et al., 2016; Vetter et al., 2013) and greater intensity of pain (Gauntlett-Gilbert & Eccleston, 2007; Guite et al., 2007; Kashikar-Zuck et al., 2001; Konijnenberg et al., 2005; Sil et al., 2016) experience more functional disability. In youth with chronic idiopathic pain, Kashikar-Zuck et al. (2001) observed that pain intensity accounted for 58% of the variance in functional disability. Therefore, it is well-documented in the literature that pain frequency and intensity significantly predicts youth functional disability. Relatedly, greater pain frequency (Rabbitts et al., 2005; Palermo, Harrison

& Koh, 2006; Barakat et al., 2008) and intensity (Rabbitts et al., 2015; Huguet & Miro, 2008) have also been documented to predict worse HRQoL. In a sample of school-age children, Petersen et al. (2009) observed that average HRQoL scores were lower, indicative of worse HRQoL, in youth with chronic pain than in youth without chronic pain. Pain characteristics have also been documented to be significantly associated and predict other health-related outcomes (e.g., internalizing symptoms; Ramchandani et al., 2007).

1.3.1 Pain Catastrophizing

Although there is literature suggesting that pain frequency and intensity are significantly associated with health-related outcomes, some researchers argue that it is not the experience of pain, but rather, the evaluative component or appraisal of pain that is critical (e.g., Miller et al., 2018). Pain catastrophizing, the tendency to magnify the threat value of a pain stimulus; to feel helpless in the context of pain; and to be unable to inhibit pain-related thoughts in anticipation of, during, or following a painful encounter is suspected to play a significant role in youth's health-related outcomes (Pielech et al., 2014; Jastrowski Mano et al., 2011; Quartana et al., 2009; Sil et al., 2016). Despite moderate correlations with anxiety, it is broadly accepted that this cognitive appraisal of pain is, and has been demonstrated to be, statistically and conceptually distinct from anxiety; moreover, pain catastrophizing appears to more strongly predict functional outcomes respective to anxiety (Benore et al., 2015; Eccleston et al., 2004; Tran et al., 2015). Pain catastrophizing may be conceptualized as either trait or state pain-specific worries (Durand et al., 2017). In a community sample of children, who underwent a pain induction task (e.g., cold-pressor task, heat pain simulation, pressure pain), state pain catastrophizing appeared to have a stronger contribution to pain-related outcomes (e.g., pain intensity) when compared to trait pain catastrophizing (Durand et al., 2017). However, most pain catastrophizing assessment

measures examine trait pain catastrophizing (e.g., a child's general tendency to catastrophize about pain), which have demonstrated that high levels of trait pain catastrophizing predict worse health-related outcomes for youth with chronic diseases who experience pain (Sil et al., 2016; Cunningham et al., 2014).

Previous literature in pediatric chronic pain populations has documented the predictive role of pain catastrophizing on pain frequency and intensity as well as other health-related outcomes including functional disability and HRQoL (Cunningham et al., 2014; Levy et al., 2014; Lynch-Jordan et al., 2013; Pielech et al., 2014; Tran et al., 2015). Specifically, the literature suggests that greater youth pain catastrophizing is associated with greater functional disability (Cunningham et al., 2014; Lynch-Jordan et al., 2013; Miller et al., 2018; Tran et al., 2015; Weiss et al., 2013). Miller et al. (2018) conducted a meta-analysis of studies that examined pain, pain catastrophizing, and functional outcomes in youth with pain; they noted a moderate association between pain catastrophizing and functional disability. The negative impact of pain catastrophizing and its positive relationship with functional disability has been documented in predominantly pediatric chronic pain samples, including youth with functional abdominal pain, chronic pain, and irritable bowel disease (Langer et al., 2009; Vervoort et al., 2006; Wojtowicz et al., 2013).

It has been documented across pediatric chronic pain populations that youth pain catastrophizing is associated with worse HRQoL. Specifically, studies have found negative associations between youth pain catastrophizing and HRQoL across youth with chronic pain (Tran et al., 2015; Cousins et al., 2015), organic gastrointestinal disorders (Warschburger et al., 2014), and irritable bowel disease (De Carlo et al., 2019). Together, these data suggest that as youth pain catastrophizing increases, youths' report of HRQoL decreases, indicating worse

HRQoL. These investigations document efforts to expand the literature that examines the detrimental impact of pain catastrophizing on functional and psychosocial outcomes in youth with chronic illnesses.

1.4 Pain Catastrophizing in Pediatric Sickle Cell Disease

A handful of studies have investigated pain catastrophizing in individuals with SCD (Citero et al., 2007; Finan et al., 2018; Mathur et al., 2016), and findings suggest that individuals with SCD catastrophize more than individuals with other chronic illnesses or healthy controls (Hollins et al., 2012; Mathur et al., 2016). Mathur et al. (2016) assessed whether catastrophizing varies by context, specifically examining catastrophizing in the context of SCD-specific pain, non-SCD pain, and laboratory pain; findings suggest that individuals reported greater catastrophizing about SCD-specific pain (Mathur et al., 2016). To date, there have only been two studies that have examined pain catastrophizing in children with SCD (Bakshi et al., 2018; Sil et al., 2016). Bakshi et al. documented that youth with SCD reported significantly greater levels of pain catastrophizing than healthy controls (Bakshi et al., 2018). Given the limited pediatric SCD literature, it is difficult to compare catastrophizing in youth with SCD to youth with chronic pain. However, a comparison of mean levels of catastrophizing as measured by the Pain Catastrophizing Scale-Child (PCS-C; Crombez et al., 2003) – the most commonly used measure of pediatric catastrophizing – suggests that youth with SCD report similar or higher levels of catastrophizing when compared youth with chronic pain (Bakshi et al., 2018; Cunningham et al., 2014; Lynch-Jordan et al., 2013; Sil et al., 2016; Tran et al., 2015). To date, there have only been two studies that have examined pain catastrophizing and health-related outcomes in pediatric SCD. Sil et al. (2016) were the first to examine pain catastrophizing in youth with SCD. The authors included 100 8- to 18-year-olds with SCD. Findings revealed that pain catastrophizing partially mediated the relationship between pain and functional disability. Bakshi et al. (2018)

were the first group to examine the relationship between child pain catastrophizing and HRQoL in a pediatric SCD sample. Participants included 33 youth with SCD and 27 healthy controls, between the ages of 8 and 21. Findings revealed an inverse correlation between child pain catastrophizing and HRQoL, only in youth with SCD (Bakshi et al., 2018).

1.5 Parent Response to Pain and Pain Catastrophizing

The literature has emphasized the key role that parents play in pediatric pain (e.g., Palermo & Chambers, 2005; Palermo, Valrie, & Karlson, 2014). The impact of parent response to youth pain is illustrated by a theoretical model proposed by Palermo and Chambers (2005). This model emphasizes a range of individual and family contextual variables that influence the relationship between children's pain and outcomes, and specifically argues that parents' responses and children's pain behavior should be considered as interacting in a reciprocal fashion.

The most commonly used measures of parent responding to pediatric pain is the Adult Responses to Children's Symptoms (ARCS; Walker & Greene, 1991). Factor analyses have suggested that the ARCS includes three factors: Protect, Minimize, and Encourage/Monitor (Claar et al., 2010; Van Slyke & Walker, 2006). The Protect factor describes parent protective, care-taking, or solicitous behaviors, which may be similar to overprotective parenting, in which parents control children's behaviors and increase comfort, inherently limiting children's opportunities to cope with stress independently (Kiel & Maack, 2012; Ungar, 2009). The Minimize factor describes parent behaviors that minimize children's pain or criticize it as excessive, which may be similar to parent responsiveness, a key dimension of parenting style (Spera, 2005), and the Encourage/Monitor describes parent behavior that encourages children to continue in daily activities while monitoring symptoms, consistent with broad surveillance

parenting practices to monitor child activities (e.g., after-school activities, homework completion, etc.) (Spera, 2005; Van Slyke & Walker, 2006).

The literature suggests that parent protecting or solicitous responding to children's pain is associated with higher levels of youth's sick-role behaviors (Logan, Simons, & Carpino, 2012). This parent response style is also associated with increased pain duration or somatic symptoms, greater number of healthcare visits for somatic symptoms, increased anxiety, increased depression, increased number of school absences, decreased school functioning, increased functional disability (Claar et al., 2010; Langer et al., 2009; Logan, Simons, & Carpino, 2012; Sieberg, Williams, & Simons, 2011; Walker, Levy, & Whitehead, 2006; Van Slyke & Walker, 2006). In addition, parent solicitous behavior reinforces children's maladaptive, short-term goals (e.g., increased attention and special privileges, decreased responsibilities, school absences). Parent protective behavior may communicate parents' own anxiety about their child's pain, which may convey to anxious children that their pain is serious and requires limitation of daily activities. In fact, data suggest that this parent response style might maintain pain (Walker & Zeman, 1992; Claar et al., 2010). Respective to literature that has examined protective parent responses to child pain, few investigations have documented the negative impact of hostile and overly critical parent responses to children's pain (Claar et al., 2008; Simons, Claar, & Logan. 2008). This parent response style, in which parents minimize, criticize, or discount children's pain, is associated with an increase of somatic symptoms (Claar et al., 2008; Simons, Claar, & Logan, 2008). It is posited that harsh responding may lead children to magnify their symptoms in effort to communicate their level of distress, when their pain is ignored (Claar et al., 2008). By contrast, parent behavior that encourages children to continue with daily activities and monitor

symptoms is associated with decreased functional disability (Cunningham et al., 2014; Vervoort et al., 2011).

Of note, investigations that have examined parent response to pediatric pain have predominantly included White, affluent families. However, a few investigations have begun to examine how cultural identity may impact and contribute to differences in parent responses to pediatric pain (El-Behadli et al., 2017; Walker et al., 2006). Data suggest that parents, who identify as an ethnic minority or immigrant, may experience different emotional and behavioral responses to children's pain and employ different stress coping mechanisms (Batista et al., 2012; Jimenez et al., 2010). El-Behadli et al. (2017) used the ARCS to compare parents of White children, Black children, Hispanic children, and multiracial children, ages 8-17 who were receiving treatment at a chronic pain specialty clinic. Findings suggested that parents of Black and Hispanic children reported more protectiveness and monitoring of children's pain, respective to that reported of parents of White children (El-Behadli et al., 2017). This is consistent with findings from Walker et al. (2006), which suggest that parents, who identify as an ethnic minority, reported increased protectiveness. It has been hypothesized that parents, who identify as an ethnic minority, may engage in more protectiveness, in response to increased challenges, stress, and discrimination in medical settings (Meyer, 2003; Mougianis et al., 2020). Therefore, it is possible that Black parents of children with SCD may respond to SCD-pain differently than the predominately White parents of children with non-SCD chronic pain.

Thus far, a handful of investigations have examined the interaction between parents' response to children's pain catastrophizing or children's emotional distress related to pain. Vervoort et al. (2011) conducted a cross-sectional study with Flemish school children between 8 and 18 years of age and their parents. Findings revealed that parent response to child pain

moderated the relationship between child pain catastrophizing and functional disability, such that, protecting or solicitous parent responses to child pain strengthened the relationship between child pain catastrophizing and functional disability. Additionally, analyses suggested that parent promotion of adaptive coping in children buffered against the detrimental impact of pain catastrophizing on children's functional abilities, thereby decrease functional disability (Vervoort et al., 2011). A study conducted by Claar et al. (2008) examined the relationship between children's emotional distress, parent response to child pain, and children's functional disability and somatic symptoms in children between 8 and 17 years of age with chronic pain. Analyses were stratified by three different profiles of parent responses, specifically protectiveness, minimization, and encouraging and monitoring of child pain symptoms. Findings revealed that child anxiety moderated the relationship between parent response to child functional disability, such that at high levels of anxiety, the relationship between parent engagement in protective responses and functional disability was strengthened (Claar et al., 2008). Researchers also examined whether children's emotional distress moderates the relationship between parent response to child pain and functional disability in youth with headache, juvenile idiopathic arthritis, and SCD between 8 and 16 years of age (Peterson & Palermo, 2004). Findings revealed that for children with higher levels of emotional distress and greater anxiety and depressive symptoms, the relationship between parent engagement in solicitous responses to child pain and functional disability was strengthened (Peterson & Palermo, 2004). Together, these studies suggest that more anxious children, or children who engage in more pain catastrophizing, appear to be particularly more vulnerable to the negative impact of parental protectiveness.

Studies suggest that children's internal experience of pain interacts with parents' response to children's pain symptoms to predict functional outcomes. There have been a few

investigations that have sought to examine the relationship between these child and parent factors in youth with chronic pain in the context of meditation analyses (Guite et al., 2011; Cunningham et al., 2014; Welkom et al., 2013); however, these analyses fail to take into account the interactional nature of child pain catastrophizing and parent response to child pain (e.g., Palermo & Chambers, 2005). In sum, the literature generally highlights the negative impact of parent protective and minimizing behavior and the value of engaging in encouraging and monitoring behavior; however, no study to date has examined the interaction of child catastrophizing and parent responding on child HRQoL in pediatric SCD. Additionally, no studies to date have examined parent response to pediatric SCD or an entirely Black sample.

1.6 Summary

Youth with SCD experience frequent pain, which is related to their functioning and HRQoL. Data suggest that rather than the pain experience itself, the appraisal of pain is the critical variable. There is a dearth of literature examining pain catastrophizing in pediatric SCD; and to date, there are no studies that examine parent factors that might exacerbate or buffer the relationship between child catastrophizing and health-related outcomes. The purpose of the current study is to examine the relationship between child pain catastrophizing, parent responses to child pain, and health-related outcomes in pediatric SCD. Moreover, I strive to examine the potential moderating impact of parent behavioral responses to youth pain symptoms on the relation between youth pain catastrophizing and youth health-related outcomes. Pain catastrophizing has been examined in other pediatric pain populations; however, no literature to date has examined the interaction of children's pain catastrophizing and parents' response to children's pain with respect to health-related outcomes in a pediatric SCD sample. My findings from this investigation could be clinically valuable, as child cognitions and parent behavior are modifiable factors that may be targeted with appropriate interventions.

1.7 Primary Aims and Hypotheses

In this study, my first aim was to understand the relationship between child pain catastrophizing and child health-related outcomes, specifically functional disability and HRQoL in a pediatric SCD sample. I hypothesized that greater pain catastrophizing would predict greater functional disability and worse HRQoL. Second, I aimed to examine the relationship between parent response to child pain symptoms and child pain catastrophizing and health-related outcomes. I hypothesized that parent response to child pain would moderate the relationship between child pain catastrophizing and health-related outcomes (i.e., functional disability, HRQoL). Specifically, I expected that at higher levels of parent protective and minimizing responses, the positive relationship between child pain catastrophizing and functional disability as well as the inverse relationship between child pain catastrophizing and HRQoL would be strengthened. In addition, I predicted that higher levels of parent encourage/monitor responses would buffer the positive relationship between child pain catastrophizing and functional disability and also the negative relationship between child pain catastrophizing and HRQoL.

2 METHOD

2.1 Participants

The proposed project is part of a larger study examining psychosocial variables in a sample of 100 youth with SCD. To determine the size of effect that may be detected with the current sample size, a sensitivity power analysis using 3 predictors was conducted with an alpha level of .05 and power of .80 using G*Power 3.1.9.4 (Faul et al., 2009). A small to moderate effect size of .11 was indicated, which is consistent with prior studies that have found similar effect sizes when examining the interaction of pain catastrophizing and parenting behavior on health outcomes (i.e., Claar et al., 2008; Vervoort et al., 2011).

Participants were 100 youth with SCD between 8 and 18 years old recruited from a large SCD clinic in the southeastern United States. To be included in the larger study, youth had to have experienced SCD-related pain in the past 30 days and speak English as their primary language. Patients were excluded if there was significant cognitive or development disabilities, which would interfere with their ability to understand survey questions.

2.2 Procedure

Participants were approached at routine outpatient clinic visits, inpatient admission, or via a mailed letter following a missed outpatient visit. Eligibility was assessed by a research coordinator. For those approached in-person, details about the study were provided and interest was assessed. Participants were then provided the opportunity to have all questions answered before deciding about study participation. Parents or legal guardians provided written consent for their child to participate in the study, and youth gave written assent. If the parent was not present at the time of recruitment, verbal consent was obtained by phone. All participants were given a copy of the document for their records. The participant had the option to participate at their next routine clinic visit, to complete questionnaires online via a secure website, or to complete a packet of questionnaires at home. If participants preferred to complete questionnaires at home, appropriate directions for electronic access or a packet of questionnaires were provided. For those mailed a letter, a member of the research team followed up by phone, one week later, to describe the study and assess interest in participation. After the participants provided consent and assent, participants were asked to complete a battery of questionnaires about their pain and psychosocial functioning related to SCD, which required approximately 30-45 minutes. Parent-child dyads were paid $30 to compensate them for their time and efforts. At the end of study

participation, an informal debriefing occurred, during which participants were provided the opportunity to ask questions about their participation and the larger investigation.

2.3 Measures

Youth and parent participants completed separate self-report measures. For the current study, youth completed questionnaires that assessed pain frequency, pain catastrophizing, functional disability, and health-related quality of life. Parents completed questionnaires that assessed demographic characteristics and parent response to child pain.

2.3.1 Demographics (Appendix A)

Demographic data were collected using a demographic measure to assess information about the parent (i.e., age, gender, race, health status, parent education, and family income) and the child (i.e., age, gender, race, health history, and health status).

2.3.2 Pain Frequency (Appendix B)

Pain frequency was measured by child self-report of the number of days with pain in the past month. While this approach to pain assessment has been critiqued as limited in scope and unable to identify confounding factors or affective contributions to pain (Cohen et al., 2020), this measurement was utilized to be consistent with prior pediatric pain literature and allow for comparison across investigations (Tran et al., 2015; Bakshi et al., 2017).

2.3.3 Youth Pain Catastrophizing (Appendix C)

Youth pain catastrophizing was measured via the child self-report Pain Catastrophizing Scale-Child (PCS-C; Crombez et al., 2003). The PCS-C is a 13-item self-report questionnaire that assesses thoughts and feelings about pain. Each item is rated on a 5-point Likert scale from *mildly* (0) to *extremely* (4). Total scores range 0-52, with higher scores indicative of greater catastrophic thinking in response to pain. The scale has commonly been used in various pediatric

chronic pain populations and is well-validated. High internal reliability for the PCS-C was found in a prior study (α=0.87; Crombez et al., 2003). The internal consistency in the current sample was 0.92, indicating strong internal consistency.

2.3.4 Parent Response to Youth Pain (Appendix D)

Parent response to youth's pain was measured by the parent-completed Adults' Response to Children's Symptoms (ARCS; Van Slyke & Walker, 2006). The ARCS is a 29-item self-report questionnaire that assesses parents' responses to their child's or adolescent's pain symptoms. Each item is rated on a 5-point Likert scale from *never* (0) to *always* (4). Item responses are summed to create three subscale scores – Protect, Minimize, and Encourage/Monitor. This measure has been well-validated in pediatric populations and demonstrates moderately high reliability measures (Protect; α=0.86; Minimize; α=0.79; Encourage/Monitor: α=0.83; Claar et al., 2010). The Protect, Minimize, and Encourage/Monitor subscale scores were used for analyses in the current study, and Cronbach's alphas were 0.89, 0.77 0.65 respectively, suggesting adequate internal consistency in the study sample.

2.3.5 Functional Disability (Appendix E)

Youth functional disability was measured with the Functional Disability Inventory (FDI; Walker & Greene, 1991). The FDI is a 15-item self-report questionnaire that assesses youth's perceived difficulty to perform daily activities at home, school, and recreational/social settings. Each item is rated on a 5-point Likert scale (0 = *no trouble* to 4 = *impossible*). The range of scores are 0 to 60, with higher scores indicative of greater functional disability. This measure is widely used across studies with samples of children with chronic pain and is well-validated. Internal reliability for the FDI has been found to be high (α=0.86-0.91; Claar & Walker, 2006) and was 0.92 in the current sample.

2.3.6 Health-Related Quality of Life (Appendix F)

Youth's perception of health-related quality of life was measured by the Pediatric Quality of Life Inventory – Sickle Cell Module (PedsQL-SCD; Panepinto et al., 2012). The PedsQL-SCD is a 43-item questionnaire that assesses various domains of quality of life, including Pain and Hurt, Pain Impact, Pain Management and Control, Worry I, Worry II, Emotions, Treatment, Communication, and Communication II. Each item is rated on a 5-point Likert scale (0 = *never a problem* to 4 = *always a problem*). Items are reverse-scored and transformed to a 0-100 scale, such that higher scores indicate better health-related quality of life. Internal reliability for the PedsQL Child Self-Report in youth with SCD has been found to be high (α=0.90; Panepinto et al., 2013). Total scores were utilized in analyses, and internal consistency was 0.96, suggesting strong internal validity.

3 RESULTS

3.1 Preliminary Analyses

Sample descriptive statistics, including frequencies, means, and standard deviations were calculated to characterize the sample (i.e., child age, child sex, child race, SCD genotype, parent age, parent sex, parent race, marital status, relationship to child, highest level of education achieved, number of children, annual household income) (Tables 1 and 2). One parent did not provide parent age or number of children, and seven parents did not report annual income. Subsequently, frequencies, means, and standard deviations of the study variables were calculated (i.e., disease severity, pain frequency, child pain catastrophizing, parent protectiveness, parent minimization, parent encouragement/monitoring, functional disability, child health-related quality of life) (Table 3).

Table 1. Child Participant Demographic Information (N=100)

Variable	***M (SD)***
Age, mean (SD)	13.53 (2.73)
Variable	**N (%)**
Sex	
Male	40 (40%)
Female	60 (60%)
Ethnicity	
Hispanic/Latino	2 (2%)
Not Hispanic/Latino	98 (98%)
Race	
Black	94 (94%)
Biracial/multiracial	2 (2%)
Decline	4 (4%)
Sickle Cell Disease Genotype	
HbSS	75 (75%)
HbSC	16 (16%)
SB^{+}Thal	5 (5%)
SB^{0}Thal	3 (3%)
Not sure	1 (1%)

Table 2. Parent Participant Demographic Information (N=100)

Variable	***M (SD)***
Age	41.82 (6.54)
Number of children	2.73 (1.62)
Variable	**N (%)**
Sex	
Male	13 (13%)
Female	87 (87%)
Ethnicity	
Hispanic/Latino	1 (1%)
Not Hispanic/Latino	99 (99%)
Race	
Black	93 (93%)
American Indian or Alaskan Native	1 (1%)
Other	1 (1%)
Biracial/multiracial	2 (2%)
Decline	3 (3%)

Relationship to child	
Mother	85 (85%)
Stepmother	1 (1%)
Grandmother	1 (1%)
Father	12 (12%)
Stepfather	1 (1%)
Marital status	
Single	37 (37%)
Married	42 (42%)
Divorced	9 (9%)
Separated	8 (8%)
Widowed	4 (4%)
Highest level of education achieved	
Less than 7th grade	1 (1%)
Junior high school (9th grade)	1 (1%)
Partial high school (10th or 11th grade)	2 (2%)
High school graduate	24 (24%)
Trade school	5 (5%)
Partial college ($\geq$ 1 year specialized training)	25 (25%)
College or university graduate (BA/BS)	26 (26%)
Graduate or professional degree	16 (16%)
Number of children	
1	20 (20.2%)
2	33 (33.3%)
3	22 (22.2%)
4	15 (15.2%)
5	3 (3%)
6	3 (3%)
7	2 (2%)
11	1 (1%)
Missing	1
Annual household income	
<$4,000	8 (8%)
$4,000-$7,000	6 (6%)
$7,001-$10,000	8 (8%)
$10,001-$13,000	6 (6%)
$13,001-$16,000	8 (8%)
$16,001-$20,000	3 (3%)
$20,001-$30,000	8 (8%)
$30,001-$50,000	22 (22%)
$50,001-$75,000	12 (12%)
$\geq$ $75,000	12 (12%)
Decline	7 (7%)

Table 3. Descriptives of Outcome Study Variables

Variable	**N (%)**
Disease severity	
Mild	22 (22%)
Moderate	41 (41%)
Severe	37 (37%)
Variable (Range)	**M ± SD**
Pain frequency (days in past month) (0-31)	11.01 ± 10.08
Child pain catastrophizing (0-52)	25.25 ± 12.21
Parent protectiveness (0-3.80)	2.44 ± 0.68
Parent minimization (0-3.17)	0.97 ± 0.74
Parent encouragement/monitoring (1.46-1.50)*	1.47 ± 0.01
Functional disability (0-42)	15.55 ± 11.52
Health-related quality of life (13.95-100)	53.85 ± 18.76

**** One outlier corrected, and variable received log[10] transformation***

Pearson correlations were conducted to examine relationships between demographic variables (e.g., child age, parent age, number of children) and study variables (Table 4). Analyses revealed that child age was positively correlated with parent age (r = .31, p < .01), pain frequency (r = .38, p < .01), functional disability (r = .20, p = .04), and negatively correlated with parent protectiveness (r = -.23, p = .02) and health-related quality of life (r = -.23, p = .02). Parent age was positively correlated with annual household income (r = .30, p < .01) and negatively related to parent protectiveness (r = -.23, p = .02). Annual household income was positively correlated with parent education (r = .51, p < .01) and health-related quality of life (r = .21, p = .04), and was negatively correlated with pain catastrophizing (r = -.29, p < .01) and parent protectiveness (r = -.26, p = .01). Parent education was positively associated with functional disability (r = .22, p = .03) and negatively associated with parent minimization (r = -.27, p < .01). Correlations revealed that disease severity was positively associated with pain frequency (r = .38, p < .01), pain catastrophizing (r = .22, p = .03), parent protectiveness (r = .24, p = .02), and functional disability (r = .27, p < .01), and negatively associated with health-related quality of life (r = -.34, p < .01). Number of children was not significantly correlated with

demographic or primary outcome variables. Pearson correlations were also conducted to explore associations among study variables (Table 4). Analyses revealed that child pain catastrophizing was positively correlated with pain frequency (r = .28 , p < .01), parent protectiveness (r = .20, p = .04), parent minimization (r = .34, p < .01), functional disability (r = .31, p < .01) and negatively correlated with parent encouragement/monitoring (r = -.32, p < .01) and health-related quality of life (r = -.60, p < .01). Parent protectiveness was negatively correlated with parent encouragement/distraction (r = -.59, p < .01). Health-related quality of life was negatively correlated with pain frequency (r = -.51, p < .01), parent minimization (r = -.25, p = .01) and functional disability (r = -.47, p < .01), and positively correlated with parent encouragement/monitoring (r = .23, p = .03).

Table 4. Intercorrelations Among Study Variables

Variable	1	2	3	4	5	6	7	8	9	10	11	12	13
1. Child age	1.00												
2. Parent age	.31**	1.00											
3. Number of children	.07	.13	1.00										
4. Parent education	.03	.18	-.02	1.00									
5. Annual household income	.04	.30**	<.01	.51**	1.00								
6. Pain frequency	.38**	.05	-.06	.11	.13	1.00							
7. Disease severity	.01	-.08	.05	.07	-.04	.38**	1.00						
8. Child pain catastrophizing	.08	-.06	-.07	-.12	-.29**	.28**	.22*	1.00					
9. Protectiveness	-.23*	-.23*	.06	-.09	-.26**	.01	.24*	.20*	1.00				
10. Minimization	.12	.10	-.04	-.27**	-.20	.15	-.05	.34**	-.05	1.00			
11. Encouragement/Monitor	.10	.16	.10	.10	.18	-.09	-.18	-.32**	-.59**	-.09	1.00		
12. Functional disability	.20*	.11	.04	.22*	.15	.35**	.27**	.31**	.17	.02	-.07	1.00	
13. Child health-related quality of life	-.23*	.08	-.03	.10	.21*	-.51**	-.34**	-.60**	-.16	-.25*	.22*	-.47**	1.00

Notes. * p < .05; ** p < .001.

An independent-samples t test revealed a statistically significant difference in functional disability between female children (M = 17.47, SD = 10.84) and male children (M = 12.68, SD = 12.04); t (98) = 2.07, p = .04; no statistically significant differences were observed between HRQoL by child sex. An independent-samples t test revealed no differences in outcomes variables (functional disability or HRQoL) between children of female and male caregivers. A one-way ANOVA revealed statistically significant differences between youth SCD genotypes for

functional disability [$F(3, 95) = 5.92$, $p < 0.01$)]; there was homogeneity of variances, as assessed by Levene's test for equality of variances (p =.621). A Tukey post hoc test revealed that youth with HbSC genotype reported significantly greater levels of disability (M = 25.63, SD = 11.26) when compared to youth with the HbSS genotype (M = 13.28, SD = 10.76, $p < 0.01$). No other statistically significant differences were observed by youth SCD genotype. A one-way ANOVA revealed statistically significant differences in functional disability between youth with mild, moderate, and severe disease [$F(2, 97) = 9.31$, $p < 0.01$)]; the assumption of homogeneity of variances was violated, as assessed by Levene's test for equality of variances (p = .019). A Games-Howell post hoc test revealed that youth with moderate disease reported significantly greater functional disability (M = 19.00, SD = 10.98) when compared to youth with mild disease (M = 7.09, SD = 8.00, $p < 0.01$). Youth with severe disease reported significantly greater functional disability (M =16.76, SD = 11.61) when compared to youth with mild disease (M = 7.09, SD = 8.00, $p < 0.01$); no statistically significant differences in functional disability were observed between youth with moderate and severe disease. Additionally, a one-way ANOVA revealed statistically significant differences in HRQoL between youth with mild, moderate, and severe disease [$F(2, 97) = 10.4$, $p < 0.01$)]; there was homogeneity of variances, as assessed by Levene's test for equality of variances (p = .437). A Tukey post hoc test revealed that youth with moderate disease reported significantly worse health HRQoL (M = 49.58, SD = 15.52) when compared to youth with mild disease (M = 68.62, SD = 20.06, $p < 0.01$). Youth with severe disease reported significantly worse HRQoL (M = 49.80, SD = 17.17) when compared to youth with mild disease (M = 68.62, SD = 20.06, $p < 0.01$); no statistically significant differences in HRQoL were observed between youth with moderate and severe disease. An independent-samples t test = revealed no statistically significant differences in outcome variables between

children with mothers and fathers as caregiver. A statistically significant difference in functional disability was observed between children whose parent was separated (M = 25.38, SD = 9.94) and children whose parent was widowed (M = 8.25, SD = 4.11, p = .01), as revealed by a one-way ANOVA [$F(4, 95) = 3.14$, p = .02)] with Games-Howell post hoc tests, following violation of assumption of homogeneity of variance assessed by Levene's test (p = .016); no statistically significant differences by caregiver marital status were observed for health-related quality of life. A one-way ANOVA revealed no statistically significant differences in outcome variables by caregiver education level or annual household income.

Based on these analyses, child age was entered as a covariate in primary analyses involving regression and moderation, in addition to pain frequency over the past thirty days and disease severity, a proxy of SCD genotype. Though analyses suggest that child sex predicted differences in only one outcome variable, the literature suggests that female sex negatively predicts differences in both functional disability (Claar & Walker, 2006; Walker & Greene, 1991) and health-related quality of life (Dampier et al, 2010; Panepinto & Bonner, 2012). Therefore, child sex was also entered as a covariate in regression and moderation analyses. Similarly, annual household income was only associated with health-related quality of life; however literature suggests that income predicts functional disability (Palermo et al., 2008) and health-related quality of life (Erickson, Munzenberger, Plante, et al., 2002; Mansour, Kotagal, Rose, et al., 2003), which prompted inclusion of income as a covariate in regression and moderation analyses.

Before conducting the regression analyses, regression assumptions (i.e., assumptions of independence, linearity, normality, homoscedasticity and outliers) were checked for violations; most regression assumptions were met (Field, 2009). The assumption of normality, linearity, and

homoscedasticity were confirmed by running scatter plots of residuals. Normality was assessed by calculating statistics for skew and kurtosis of each study variable; violations of skew were observed on the ARCS Encourage/Monitor variable. Outlier analysis was pursued by assessments of distance and leverage; boxplot analyses of each study variable facilitated outlier identification. One problematic outlier was identified on the ARCS Encourage/Monitor variable, which was corrected to be within three standard deviations of the mean. Additionally, a $\log^{10}$ transformation of the ARCS Encourage/Monitor variable was conducted to address violation of normality. Subsequently, the assumption of normality was met. While subsequent analyses revealed moderate improvement, outlier assumptions continued to be violated after outlier correction and $\log^{10}$ transformation. In effort to minimize over-manipulation of the dataset, no additional corrections were made. Collinearity statistics were examined (i.e., Tolerance and VIF) and were within acceptable limited, which suggested that the assumption of multicollinearity was met.

3.2 Primary Analyses

The first goal of analyses was to examine the unique predictive value of child pain catastrophizing on both functional disability and HRQoL, above and beyond child age, child sex, disease severity, pain frequency, and income. Therefore, two hierarchical regressions were performed. Subsequently, to test the moderating role of parent response to child pain symptoms on the relation between child pain catastrophizing and health-related outcomes (e.g., functional disability, HRQoL), predictor variables were centered. Hierarchical multiple regression analyses were conducted, and interaction effects were probed using the SPSS PROCESS macro (Hayes, 2013), which uses bootstrapping, a nonparametric resampling technique (5,000 samples) to assess effects (Preacher & Hayes, 2004). Statistically significant interactions were interpreted by

plotting regression lines for low (1 standard deviation below the mean) and high (1 standard deviation above the mean) levels of parent response to child pain symptoms.

3.2.1 Predicting Functional Disability from Covariates

A two-stage hierarchical regression was conducted to determine if the addition of child pain catastrophizing improved the prediction of functional disability over and above child age, child sex, disease severity, pain frequency, and income. See Table 5 for full details on each regression model. The model of child age, child sex, disease severity, pain frequency, and income to predict functional disability was statistically significant, $R^2 = .25$, $F(5, 87) = 5.68$, $p < .01$, adjusted $R^2 = .20$. The addition of child pain catastrophizing to the prediction of functional disability led to a statistically significant increase in R^2 of .043, $F(1, 86) = 5.15$, $p = .03$. The subsequent model of child age, child sex, disease severity, pain frequency, income, and child pain catastrophizing to predict functional disability was also statistically significant, $R^2 = .289$, $F(6, 86) = 5.82$, $p < .01$, adjusted $R^2 = .239$.

Table 5. Hierarchical Regression Analyses of Covariates and Pain Catastrophizing to Predict Functional Disability

Step	*R*	R^2	*B*	SE	*t*	*LLCI*	*ULCI*	*F*Δ	ΔR^2
Block 1	.496	.246						5.684**	.246
Child age			.095	.419	.940	-.104	.291		
Child sex			-.206	2.214	-2.17*	-.394	-.017		
Disease severity			.209	1.545	2.04*	.005	.409		
Pain frequency			.291	.126	2.65**	.074	.513		
Income			.080	.374	.846	-.109	.269		
Block 2	.537	.289						5.147*	.043
Child age			.110	.411	1.11	-.085	.301		
Child sex			-.215	2.17	-2.31*	-.398	-.030		
Disease severity			.190	1.51	1.89	-.010	.386		
Pain frequency			.217	.129	1.94	-.006	.443		
Income			.156	.389	1.58	-.040	.353		
Child pain catastrophizing			.231	.097	2.27*	.029	.439		

3.2.2 Predicting HRQoL from Covariates

A second two-stage hierarchical regression was conducted to determine if the addition of child pain catastrophizing improved the prediction of HRQoL over and above child age, child sex, disease severity, pain frequency, and income. See Table 6 for full details on each regression model. The model of child age, child sex, disease severity, pain frequency, and income to predict HRQoL was statistically significant, $R^2 = .382$, $F(5, 87) = 10.74$, $p < .01$, adjusted $R^2 = .35$ The addition of child pain catastrophizing to the prediction of HRQoL led to a statistically significant increase in R^2 of .14, $F(1, 86) = 24.90$, $p < .01$. The subsequent model of child age, child sex, disease severity, pain frequency, income, and child pain catastrophizing to predict HRQoL was also statistically significant, $R^2 = .52$, $F(6, 86) = 15.56$, $p < .01$, adjusted $R^2 = .49$.

Table 6. Hierarchical Regression Analyses of Covariates and Pain Catastrophizing to Predict HRQoL

Step	*R*	R^2	*B*	SE	*t*	*LLCI*	*ULCI*	*F*Δ	ΔR^2
Block 1	.618	.382						10.738**	.382
Child age			-.041	.606	-.444	-.214	.136		
Child sex			-.074	3.20	-.860	-.239	.095		
Disease severity			-.113	2.23	-1.21	-.288	.070		
Pain frequency			-.515	.182	-5.18**	-.703	-.313		
Income			.286	.541	3.32**	.113	.448		
Block 2	.721	.520						24.898**	.139
Child age			-.067	.538	-.826	-.220	.091		
Child sex			-.059	2.84	-.771	-.205	.091		
Disease severity			-.078	1.98	-.945	-.235	.084		
Pain frequency			-.382	.169	-4.42**	-.557	-.196		
Income			.149	.509	1.84	-.012	.304		
Child pain catastrophizing			-.417	.127	-4.99**	-.578	-.249		

Moderation Analyses

3.2.3 Parent Protectiveness as a Moderator

When examining parent protectiveness as a moderator of the relationship between child pain catastrophizing and functional disability, with child age, child sex, disease severity, pain frequency, and income as included covariates, the overall model was significant, $F(8, 84) = 4.74$, $p < .01$, and accounted for 31.1% of the variance in functional disability. However, with inclusion of covariates, neither child pain catastrophizing $\beta = .21$, $t(84) = .34$, 95% CI [.002, .415], $p = .74$, nor parent protectiveness, $\beta = .18$, $t(84) = .76$, 95% CI [-.06, .42], $p = .45$, were significantly related to functional disability; and thus, the interaction term was not significant, $\beta = .02$, $t(91) = .22$, 95% CI [-.18, .22], $p = .83$. When examining parent protectiveness as a moderator of the relationship between child pain catastrophizing and HRQoL, with child age, child sex, disease severity, pain frequency, and income as included covariates, the overall model was significant, $F(8, 84) = 12.71$, $p < .01$, and accounted for 54.8% of the variance in HRQoL. With inclusion of covariates, child pain catastrophizing, $\beta = -.43$, $t(84) = -3.50$, 95% CI [-.591, -.263], $p < .01$ significantly predicted HRQoL, but parent protectiveness, $\beta = .10$, $t(84) = -1.94$, 95% CI [-.085, .294], $p = .06$, did not; the interaction term was significant, $\beta = .18$, $t(84) = 2.24$, 95% CI [.020, .341], $p = .03$. More specifically, the relationship between child pain catastrophizing and HRQoL was strongest at low levels of parent protectiveness, less strong at medium levels of protectiveness, and least strong at high levels of protectiveness (Figure 1).

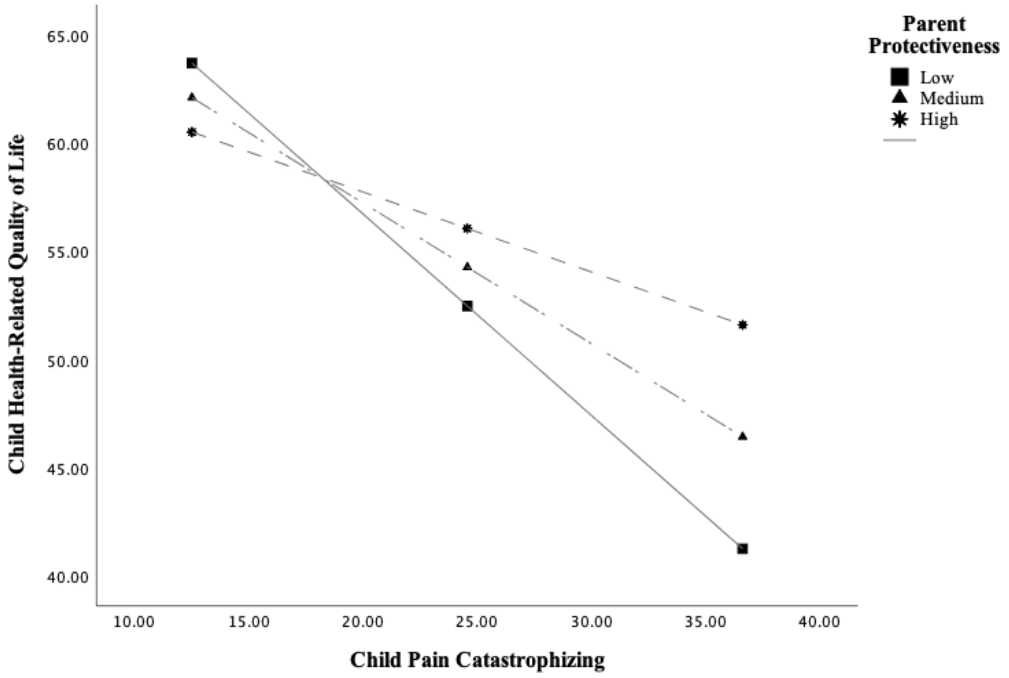

Figure 1. Parent protectiveness moderates the relationship between child pain catastrophizing and health-related quality of life.

3.2.4 Parent Minimization as a Moderator

When examining parent minimization as a moderator of the relationship between child pain catastrophizing and functional disability, with child age, child sex, disease severity, pain frequency, and income as included covariates, the overall model was significant, $F(8, 84) = 4.37$, $p < .01$, and accounted for 29.4% of the variance in functional disability. With inclusion of covariates, child pain catastrophizing, $\beta = .21$, $t(84) = 2.03$, 95% CI [-.009, .431], $p = .05$, significantly predicted functional disability but parent minimization, $\beta = .04$, t (84) = .77, 95% CI [-.170, .255], $p = .44$, did not; the interaction term was not significant, $\beta = -.07$, t (84) = -.75, 95% CI [-.251, .114], $p = .46$. When examining parent minimization as a moderator of the relationship between child pain catastrophizing and HRQoL, with child age, child sex, disease severity, pain frequency, and income as included covariates, the overall model was significant, $F(8, 84) = 13.61$, $p < .01$, and accounted for 56.5% of the variance in HRQoL. With inclusion of

covariates, both child pain catastrophizing, β = -.35, *t*(84) = -5.42, 95% CI [-.523, -.184], *p* < .01, and parent minimization, β = -.105, *t*(84) = -2.84, 95% CI [-.268, .058], *p* = .01, predicted HRQoL; the interaction term was significant, β = .20, *t*(84) = 2.85, 95% CI [.060, .340], *p* = .01. More specifically, the relationship between child pain catastrophizing and HRQoL was strongest at low levels of parent minimization, less strong at medium levels of minimization, and at high levels of minimization, the relationship between child pain catastrophizing and HRQoL was no longer statistically significant (*p* =.19) (Figure 2).

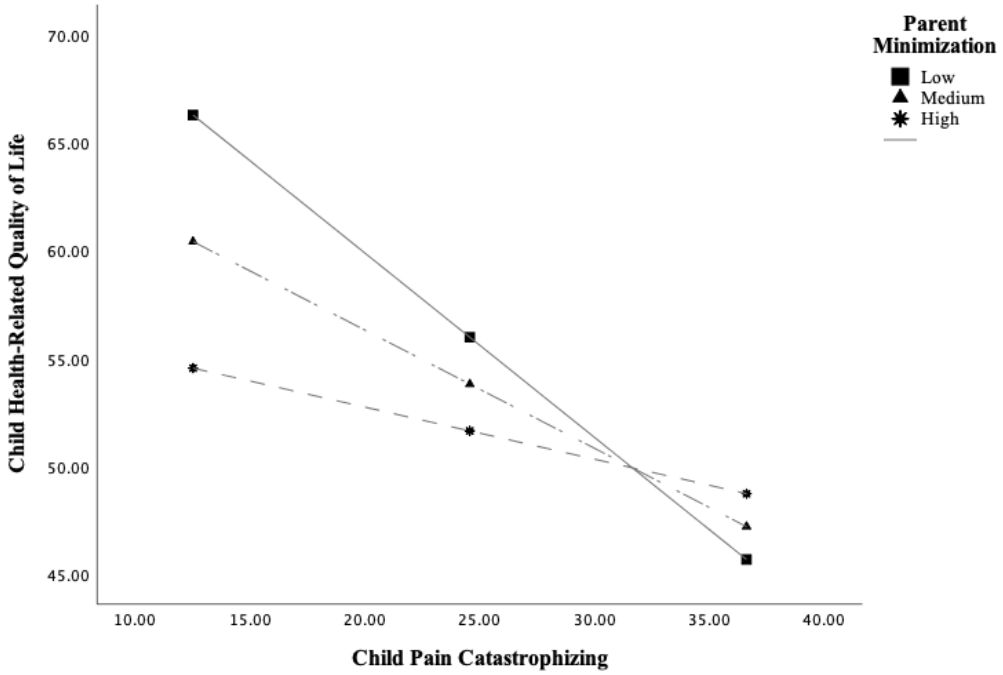

Figure 2. Parent minimization moderates the relationship between child pain catastrophizing and health-related quality of life.

3.2.5 Parent Encouragement/Monitoring as a Moderator

When examining parent encouragement/monitoring as a moderator of the relationship between child pain catastrophizing and functional disability, with child age, child sex, disease severity, pain frequency, and income as included covariates, the overall model was significant, *F*(8, 84) = 4.45, *p* < .01, and accounted for 29.8% of the variance in functional disability.

However, with inclusion of covariates, neither child pain catastrophizing, β = .27, *t*(84) = -.82, 95% CI [.045, .490], *p* = .42, nor parent encouragement/monitoring, β = -.11, *t*(84) = -.30, 95% CI [-.129, .356], *p* = .77, significantly predicted functional disability; the interaction term was not significant, β = .08, *t*(84) = .83, 95% CI [-.112, .274], *p* = .41. When examining parent encouragement/monitoring as a moderator of the relationship between child pain catastrophizing and HRQoL, with child age, child sex, disease severity, pain frequency, and income as included covariates, the overall model was significant, *F*(8, 84) = 14.99, *p* < .01, and accounted for 58.8% of the variance in HRQoL. With inclusion of covariates, child pain catastrophizing, β = -.45, *t*(84) = 3.67, 95% CI [-.621, -.287], *p* < .01, and parent encouragement/monitoring, β = -.19, *t*(84) = 2.77, 95% CI [-.367, -.003], *p* < .01, predicted HRQoL; the interaction term was significant, β = -.27, *t*(84) = -3.70, 95% CI [-.414, -.125], *p* < .01. More specifically, the relationship between child pain catastrophizing and HRQoL was strongest at high levels of parent encouragement/monitoring, less strong at medium levels of parent encouragement/monitoring, and was no longer statistically significant at low levels of parent encouragement/monitoring (*p* = .08) (Figure 3).

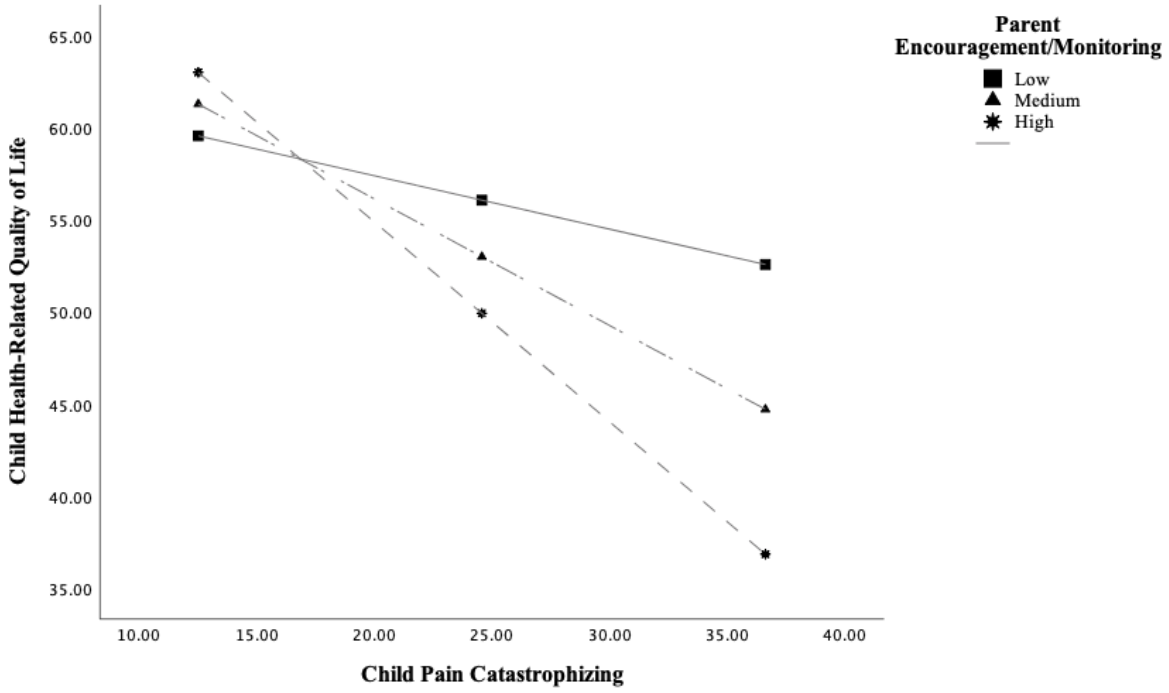

Figure 3. Parent encouragement/monitoring moderates the relationship between child pain catastrophizing and health-related quality of life.

4 DISCUSSION

Youth with SCD experience recurrent, unexpected pain, which has been shown to be associated with poor functioning and health-related quality of life (e.g., Oliver-Carpenter et al., 2011; Palermo et al., 2008; Peterson & Palermo, 2004; Sil et al., 2016; Walker & Greene, 1991). Emergent findings in the pediatric chronic pain literature suggest that youth's appraisal of pain, rather than the pain experience itself, may predict negative health-related outcomes (Pielech et al., 2014; Sil et al., 2016). However, few investigations have examined pain catastrophizing in pediatric SCD, and there are no studies to date that have identified parent factors that may exacerbate or buffer the relationship between pain catastrophizing and health-related outcomes. Thus, the aims of this study were to examine whether pain catastrophizing predicts health-related

outcomes in youth with SCD and whether parent responses to children's pain may influence that relationship.

4.1 Preliminary Analyses

With regard to the sample, the mean age of youth included was 14 years, and more female children were included than male children. On average, youth in the sample reported experiencing pain on 11 of the past thirty days. Consistent with prior pediatric SCD literature, the most prevalent SCD genotypes included were HbSS and HbSC (Bakshi et al., 2017; Fisak et al., 2011). When examining primary variables, all measures demonstrated good internal validity. Child pain catastrophizing scores were slightly lower than those found in another sample of youth with SCD (Bakshi et al., 2018), and similar (Tran et al., 2015) or slightly higher (Pielech et al., 2014) than samples of youth with chronic pain. Parent responses (protectiveness, minimization, and encouragement/monitoring) to youth's pain have not yet been examined in pediatric SCD samples. Parent protectiveness scores in the current sample were higher than those reported in pediatric chronic pain samples (Claar et al., 2008; Cunningham et al., 2014; Langer et al., 2009; Sieberg et al., 2011; Van Slyke & Walker, 2006). It is possible that this observed difference in parent response might be explained by differences in disease pathophysiology, as SCD pain is qualitatively different from non-SCD chronic pain. Given that SCD pain can indicate medical complications that warrant parent intervention or medical attention, parents of youth with SCD may engage in more protective responses than parents of children with non-SCD pain. This is different from youth who experience chronic pain, which often lacks a known pathophysiologic cause; therefore, this chronic pain does not typically require parent or medical provider intervention. These data are also consistent with prior findings that suggest that Black parents may engage in more protective behaviors respective to White parents, which has been hypothesized to be in response to increased challenges, stress, and discrimination in medical

settings (El-Behadli et al., 2017; Walker et al., 2006). Parent minimization scores were similar to those reported in parents of youth with chronic pain (Van Slyke & Walker, 2006; Claar et al., 2008; Cunningham et al., 2014). Prior to outlier correction and $\log^{10}$ transformation, parent encouragement/monitoring scores were higher than those reported in pediatric chronic pain samples (Claar et al., 2008; Cunningham et al., 2014; Van Slyke & Walker, 2006). High levels of parent encouragement/monitoring in the current sample may reflect that this is a common parent caretaking strategy used when youth experience SCD pain; Hildenbrand et al. (2015) observed that all parents included in their SCD sample reported engaging their child via encouragement/monitoring while their child experienced SCD pain. Functional disability reported in the current sample was similar to other pediatric SCD samples (Palermo et al., 2008;Oliver-Carpenter et al., 2011); however, scores were lower than those reported in pediatric chronic pain samples (Kashikar-Zuck et al., 2001; Sieberg et al., 2011; Weiss et al., 2013). Lower functional disability scores in the current sample, respective to pediatric chronic pain samples, may reflect the difference in chronicity of pain, as the current sample included youth with a range of pain frequency, which is different than youth who experience high chronicity of pain. This discrepancy may also indicate difference in severity of pain between samples and therefore lower levels of impairment. Given that youth with SCD are typically diagnosed at birth, or very early in life, youth with SCD may also have more time to adapt and develop coping strategies; therefore, they may be less functionally impaired than youth with pediatric chronic pain. Youth in the current sample reported HRQoL scores lower than other pediatric SCD samples (Bakshi et al., 2017; Panepinto et al., 2017). Discrepancy may be explained by inclusion of youth hospitalized for pain crises, which has been shown to lower report of HRQoL (Panepinto et al., 2017).

With respect to health-related outcomes, variable associations with functional disability and health-related quality of life were in the expected directions. Child age, parent education, pain frequency, disease severity, and pain catastrophizing were significantly associated with functional disability, consistent with pediatric chronic pain findings (Cunningham et al., 2014; Lynch-Jordan et al., 2013; Miller et al., 2018; Tran et al., 2015; Weiss et al., 2013). These variables, excluding parent education, and including income, parent minimization, parent encouragement/monitoring, and functional disability, were associated with HRQoL.

Protective parent responses were significantly associated with child age and parent age, such that, as children's and parents' age decreased, parent protectiveness increased. Protective parent responses were also negatively associated with annual household income, such that as income decreased, parent protectiveness increased. Disease severity and child pain catastrophizing were positively associated with parent protectiveness. Interestingly, parent encouragement/distraction was negatively associated with parent protectiveness, discrepant from reported positive association in pediatric chronic pain samples (Claar et al., 2010). This may reflect differences in patterns of parent responses to youth's pain across pediatric chronic illness populations. Pain catastrophizing was inversely associated with income, such that as income decreased, pain catastrophizing increased. Pain catastrophizing was also significantly associated with pain frequency, disease severity, parent protectiveness, parent minimization, and parent distraction. Pain catastrophizing was also positively related to functional disability and negatively related to HRQoL. Pain frequency was associated with child age, child pain catastrophizing, disease severity, and functional disability, and HRQoL.

Parents' responses to youth's pain were not significantly related to functional disability. More specifically, parent protectiveness was not related to pain frequency, functional disability,

or HRQoL. This is notably discrepant from pediatric chronic pain literature, which has documented a relationship between parent protectiveness and worse child psychosocial and functional outcomes (Claar et al., 2010; Langer et al., 2009; Logan et al., 2011; Sieberg et al., 2011; Walker et al., 2006; Van Slyke & Walker, 2006). This discrepancy may reflect that protective parent responses do not significantly impact functional disability or HRQoL for children with SCD pain, which is qualitatively different than youth with chronic pain. Parent minimization was not related to child age, parent age, pain frequency, parent protectiveness, or parent encouragement/monitoring. Parent encouragement/monitoring was not related to child age, parent age, or pain frequency. Parent age was not related to HRQoL. It is challenging to make sense of these null findings. Given that prior researchers have found significant relations among parent response to child pain symptoms and functional disability (Claar et al., 2010; Langer et al., 2009; Logan et al., 2012; Sieberg et al., 2011; Walker et al., 2006; Van Slyke & Walker, 2006), it could be that the sample was not large enough to provide the necessary power to detect relations. On the other hand, parent response may impact other psychosocial and functional outcomes that are not captured in functional disability or HRQoL. Given the limited research of parenting responses to pain in SCD samples, additional research is warranted.

4.2 Primary Analyses

Consistent with hypotheses, pain catastrophizing significantly predicted functional disability above and beyond covariates (child age, child sex, pain frequency, disease severity, income). As expected, higher levels of pain catastrophizing predicted significantly worse functional disability. This is consistent with findings observed across pediatric chronic illness populations including SCD (Sil et al., 2016), chronic pain (Vervoort et al., 2006), and inflammatory bowel disease (IBD; Wojtowicz et al., 2014). Similarly, findings suggested that pain catastrophizing significantly predicted HRQoL above and beyond covariates, such that

increased pain catastrophizing predicted significantly worse HRQoL. This observed relationship aligns with findings from pediatric chronic pain (Tran et al., 2015; Cousins et al., 2015), IBD (De Carlo et al., 2019), and other gastrointestinal disorders (Warschburger et al., 2014). Of note, these findings underscore the potential impact of youth's appraisal of pain on health-related outcomes, above and beyond their experience of pain. This investigation replicates Sil et al.'s (2016) finding that youth pain catastrophizing significantly predicts functional disability; however, this is the first study, to date, that has observed that youth's appraisal of pain appears to predict worse health-related outcomes above and beyond their experience of pain.

With respect to moderation analyses, data supported three hypothesized models; an interaction was observed between pain catastrophizing and parent protectiveness to predict HRQoL. However, directionality of the proposed interaction was the reverse of what was hypothesized; findings suggested that higher levels of parent protectiveness buffered the negative relationship between child pain catastrophizing and HRQoL. Data suggest that at low levels of pain catastrophizing, a negative cognitive response to pain, parent protective responses do not appear to significantly alter HRQoL; however, at moderate to high levels of pain catastrophizing, parent protective responses may be respectively more impactful on HRQoL. Therefore, for youth who engage in moderate to high levels of pain catastrophizing, it appears that higher levels of protective parent responses is associated with higher HRQoL. This is discrepant from what has been documented within the pediatric chronic pain literature, which has shown that parent protective responses are associated with worse health-related outcomes response (Claar et al., 2010; Langer et al., 2009; Logan et al., 2011; Sieberg et al., 2011; Walker et al., 2006; Van Slyke & Walker, 2006); however, no studies within pediatric chronic pain have examined the interaction of child pain catastrophizing and parent response to pain symptoms and therefore

assessed possible conditional effects of this parent response. This raises the question of whether this parent response is adaptive for this pediatric population, in the context of high levels of child pain catastrophizing. Interestingly, no significant correlation between parent protective responses and HRQoL was observed, which may underscore the conditional effect of this parent response on youth's HRQoL. It is possible that children, who engage in high levels of pain catastrophizing, lack alternative coping skills. Therefore, in the context of pain and high distress, parent attention and comfort is sought; when parent attention and comfort is given , it is possible that child distress decreases, which may explain association with higher HRQoL. Observed differences may also reflect differences in pathophysiology, as SCD pain is qualitatively different from non-SCD chronic pain and increased adult attention, which allows for intervention, may be adaptive. Protective parent responses, which include taking the child to the doctor and keeping the child inside, may be adaptive for children who experience pain crises that warrant parent intervention or medical attention to treat symptoms. Similarly, this parent response may be related to the need to advocate for pain management in medical settings, in response to racial discrimination and healthcare inequities for Black children (Haywood et al., 2014; Sanders-Phillips, Settles-Reaves, Walker et al., 2009).

A second interaction was observed between pain catastrophizing and parent minimization to predict HRQoL. Directionality of the proposed interaction was the reverse of what was hypothesized; findings suggest that higher levels of parent minimization buffered the negative relationship between pain catastrophizing and HRQoL. This raises question about whether this parenting response is adaptive in the context of child pain catastrophizing in pediatric SCD; however, correlational analyses show that parent minimization is negatively associated with HRQoL, such that as parent minimization increases, youth HRQoL decreases. This negative

association is consistent with pediatric chronic pain literature, which suggests that parent minimization is associated with negative health-related outcomes, particularly pain symptom maintenance (Rook & Gauntlett-Gilbert, 2016), and has been suspected to prompt emotional and psychological distress in children (Claar et al., 2008; Rook & Gauntlett-Gilbert, 2016; Simons et al., 2008). Data from the current study suggest that the strength of the relationship between pain catastrophizing and HRQoL changes at different levels of parent minimization, which may occur because this parent response is a maladaptive response to a maladaptive cognitive appraisal of pain. The relationship between pain catastrophizing and HRQoL is strongest at low levels of parent minimization, which may highlight the strong negative impact of this cognitive appraisal on HRQoL. However, when parent minimization, a negative behavioral response to pain, increases, pain catastrophizing may contribute less variance in HRQoL because the negative behavioral response from parents may contribute respectively more variance, therefore explaining the observed decreased strength and null effect of pain catastrophizing on HRQoL at medium and high levels of parent minimizing. Analyses from this model suggest that, despite low levels of parent minimization, pain catastrophizing still negatively predicts HRQoL, which underscores the negative impact of this cognitive appraisal on youth with SCD's HRQoL. Given observed negative association between parent minimization and HRQoL, decreased strength of relationship between pain catastrophizing and HRQoL likely suggests that parent minimization also negatively impacts HRQoL.

A third interaction was observed between pain catastrophizing and parent encouragement/monitoring to predict HRQoL. Directionality of the proposed interaction was the reverse of what was hypothesized; findings revealed that at higher levels of parent encouragement/monitoring, the negative impact of pain catastrophizing on HRQoL was

strengthened. However, correlational analyses indicated a positive association between parent encouragement/monitoring and HRQoL, such that as parents engage in more encouragement/monitoring, HRQoL improves; this suggests that this parent response may also display conditional effects, with respect to contribution to positive health-related outcomes. Findings from correlational analyses are consistent with pediatric chronic pain literature, which suggests that parent encouragement/monitoring is associated with positive health-related outcomes (e.g., functional disability) (Cunningham et al., 2014; Vervoort et al., 2011). The observed interaction suggests that strength of the relationship between child pain catastrophizing and HRQoL may change at different levels of parent encouragement/monitoring, and at low levels of encouragement/monitoring, the relationship between child pain catastrophizing and HRQoL is no longer statistically significant. One possible interpretation of this interaction is that for youth who engage in low levels of pain catastrophizing, parent encouragement/monitoring may not be particularly impactful on HRQoL, which is high, respective to children who engage in higher levels of pain catastrophizing. However, for youth who engage in moderate to high levels of pain catastrophizing, a negative cognitive appraisal, higher levels of parent encouragement/monitoring appears to be associated with worse HRQoL. Therefore, for youth who engage in moderate to high levels of pain catastrophizing, parent encouragement/monitoring may be a maladaptive response. It could be that this behavioral response, which encourages persistence in daily activities, exacerbates anxiety about pain. A different interpretation of this interaction could be that for children, who have parents who tend to encourage them to persist in daily activities, while monitoring their pain, pain catastrophizing has a more negative impact than for children whose parents tend not to encourage them to persist in daily activities. Given

the study design and analysis, it is not possible to determine which interpretation may best explain the observed interaction.

Surprisingly, findings did not support an interaction between pain catastrophizing and parent response (protectiveness, minimization, encouragement/monitoring) to predict functional disability. This could simply reflect that youth's functional ability is either not impacted by parent response to pain or youth persist in daily activities in the context of chronic pain. Given that youth with SCD are diagnosed early in life, it is possible that they have developed adaptive coping skills that allow them to persist in daily activities. If so, it is likely that functional disability will be limited or may reflect less impairment, respective to pediatric chronic pain populations, who experience more frequent pain than the current sample and less time since diagnosis. Distress associated efforts to overcome SCD pain may not be captured in functional disability but in other assessments of psychosocial functioning (e.g., HRQoL). It is also possible that the current sample lacks sufficient power to detect a small effect.

4.3 Limitations and Future Directions

The current study introduces novel, clinically relevant findings to the field of pediatric SCD and adds to the growing body of literature assessing pain catastrophizing in this chronic illness population; however, there are limitations to note, which have implications for future studies. With regard to the current sample, youth reported lower functional disability and HRQoL, respective to youth in other pediatric SCD samples, which may suggest that the current sample represents a sample with less functional impairment but more severe negative impact of HRQoL; therefore findings may not generalize to all youth with SCD. Additionally, the distribution of one study variable (i.e., parent encouragement/monitoring) was negatively skewed and leptokurtic, which may also limit generalizability to the pediatric SCD population. Relatedly, the current sample size may also introduce statistical limitations, specifically the ability to detect

small interaction effect sizes. Given that Claar et al. (2008) observed small interaction effect sizes, it is possible that null findings from interactions between pain catastrophizing, parent response, and functional disability are a result of lack of power to detect small effect sizes. An a priori power analysis using 3 predictors, with an alpha level of .05, power of .80, a small effect size, revealed that 550 participants would be needed to detect a small interaction effect.

With respect to study design, the cross-sectional design of the current study limits the ability to determine causal relation between study variables and temporal sequence between child variables and parent variables. Future studies should consider use of longitudinal study design to quantify causality between pain catastrophizing, parent responses to youth pain, and health-related outcomes. This study design will also shed light on parent-child dyadic interactions and provide deeper understanding of the transactional nature of youth and parent response to SCD pain.

In terms of age range, it is notable that the wide age range of the current sample includes various developmental stages, which are characteristically distinct with respect to disease management abilities and autonomy from parents. For example, an 8-year-old likely depends upon parents more heavily for guidance about management for pain crises than a 17-year-old, who has significantly more experience managing pain crises and more advanced cognitive development (e.g., ability to think about long-term consequences). Similarly, parent responses to children likely change across developmental stages, as youth become increasingly capable of disease management; this is consistent with data from the present investigation suggest that child age and protective responses are inversely correlated, suggesting that parents engage in more protective behavior for younger children. Notably, child age is positively associated with functional disability and negatively associated with HRQoL, such that as youth get older, they

report greater functional impairment and worse HRQoL, which may be reflective of greater awareness of individual differences respective to peers. It is also notable that the measure to quantify parent response to youth pain may have a different factor structure by youth age (Noel et al. 2014). It has been observed that for children (7-11 years) a 4-factor model and for adolescents (12-18 years) a 5-factor model are superior to the 3-factor model used in the literature, which suggests that parent responses to youth's pain are uniquely different by child age. Additionally, it is possible that the intensity and frequency of pain catastrophizing changes across developmental stages; as children get older, pain catastrophizing might increase in frequency, as youth develop greater awareness of their internal experience and accumulate pain crisis experiences. By contrast, it is possible that as youth develop, they hone and integrate more coping skills, which minimizes pain catastrophizing. Future investigations should strive to assess how pain catastrophizing changes across child development. Future investigations should also strive to replicate current findings with child and adolescent-specific samples, which will allow for assessment of developmentally specific parent responses to youth pain. Moreover, a longitudinal study design will allow for assessment of how parent-youth interaction around pain management and pain response changes across developmental trajectory.

Contextualization of current findings within Afro-centered literature suggests that Black parenting styles may reflect cultural values and therefore may be qualitatively different than White parenting styles. Given preliminary data from clustered profile analysis of parenting styles, it is likely that parent responses co-occur, which may have implications for parent response to pediatric SCD pain (Rious et al., 2019). Therefore, future directions of this work might include aggregating data from the ARCS subscales to reflect combinations of parent responses to youth, characteristic of Black parenting practices. However, it is worth noting that

thus far, the ARCS has been validated in predominantly White samples, and therefore identified factors may fail to represent aspects of Black parent responses. Future directions of this work include assessment of the psychometric validity of the ARCS in a Black pediatric sample and greater research on Black parenting practices respective to pain and pain crisis management.

Future directions of this work also include examination of the impact of parent response to pain on child functioning more broadly. Given the documented relationship between parent protectiveness and school attendance and academic functioning, it is suspected that parent response has implications for education and peer-relationship development (Logan et al., 2012). Parent response, which provides guidance to youth about how to respond to pain, may also have implications coping and ability to independently manage disease symptoms. For example, if a parent responds to a child with protectiveness, a child might learn to depend on their parent whenever they experience pain. It is possible that this protectiveness might also serve to model how to care for themselves. Relatedly, future studies may assess how parent responses to children's pain are related to or influenced by other parenting factors, such as parent stress, anxiety and depression, parent-child dyadic conflict, marital relationships, number of children in the home, and parent adjustment. Of note, much of the current sample included female parents and caregivers, which may limit generalizability of the findings to male caregivers. Therefore, future studies should investigate the impact of parent gender on parent response to child and examine whether parent gender interacts with child gender to predict parent behavioral response.

Future studies may also extend this work to assess other family-systems factors that may impact Black youth with SCD's experience of pain and its impact of health-related outcomes. Exploration of additional resilience and protective factors should be identified.

4.4 Summary

Findings from the current investigation underscore the negative impact of pain catastrophizing on health-related outcomes for youth with SCD, over above children's experience of pain. Moreover, analyses suggest that impact of parents' response to children's pain may be conditional to children who engage in moderate to high levels of pain catastrophizing. Data from the current study suggest that high levels of parent protective responses buffers the relationship between child pain catastrophizing and HRQoL. More specifically, for children who engage in high levels of pain catastrophizing, high levels of parent protective responses appears to be associated with higher HRQoL; therefore, parent protective responses may be adaptive when children engage in more pain catastrophizing. Data also suggest that parent minimization is negatively associated with HRQoL and higher levels of minimizing appears to buffer the negative impact of the child's negative cognitive appraisal of pain on HRQoL; however, data suggest that parent minimizing may be more negatively impactful for youth who engage in lower levels of pain catastrophizing. Analyses also suggest that parent encouragement/monitoring is positively associated with HRQoL; however, for children who engage in lower levels of pain catastrophizing, parent encouragement/monitoring does not appear to be impactful of high levels of HRQoL. For children who engage in higher levels of pain catastrophizing, encouragement/monitoring appears to negatively impact HRQoL; therefore, parent encouragement/monitoring may be a maladaptive response for children who engage in high levels of pain catastrophizing. Together, these findings suggest that for children who engage in low levels of pain catastrophizing, parent response to pain does not appear to be particularly impactful on HRQoL. However, for youth who engage in moderate to high levels of pain catastrophizing, protective parent responses are associated with better HRQoL and encouragement/monitoring parent responses are associated with worse HRQoL. It is critical to

identify youth, who engage in moderate to high levels of pain catastrophizing, and to work collaboratively with parents and caregivers to promote adaptive behavioral responses to pain, which may include protective responses. Identification of an adaptive amount of protective behaviors is critical and warrants further exploration; qualitative interviews with parents of youth with SCD may be helpful in guiding future studies.

Together, these variables (i.e., child cognition, parent behavioral response) serve as two critical modifiable targets for clinical invention. These targets are particularly amenable to Cognitive Behavioral Therapy-based (CBT) or third-wave cognitive-behavioral treatment approaches including Acceptance and Commitment Therapy (ACT) (Weiss et al., 2013; Wetherell et al., 2011). In CBT approaches, treatment prioritizes identification of maladaptive thoughts about pain, feelings, and associated pain behaviors; challenging of maladaptive thoughts and behaviors; and increased goal-directed behavior (Cohen et al., 2020). By contrast, ACT approaches prioritize development of different responses to maladaptive thoughts and sensations that create distance between negative thoughts and experiences and the individual; also central to this theoretical orientation is persistent engagement in values-based activities and psychological flexibility (Cohen et al., 2020; Pielech et al., 2017).

Across the pediatric chronic pain literature, pain catastrophizing cognitions have been treated within these broader cognitive therapeutic treatment packages, which in many investigations appear to show statistically significant reductions in this negative cognitive appraisal (Kashikar-Zuck, Sil, Lynch-Jordan et al., 2013; Weiss et al., 2013; Wetherell et al., 2011). However, some treatments have not showed reductions in pain catastrophizing (Palermo, Law, Fales, et al., 2016). In some treatments, environmental factors, specifically parent behaviors that reinforce maladaptive thoughts or pain behaviors, are targeted (Weiss et al.,

2013). Recent CBT interventions developed for youth with SCD have shown improvements in pain coping efficacy (Schatz, Schlenz, McClellan et al., 2015; Sil, Lai, Lee, et al., 2020); however, greater exploration of treatment components that promote reduction in pain catastrophizing and parent behavioral responses is needed.

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Appendix A. Background Information

BACKGROUND INFORMATION

Instructions: We would like to ask you a few questions about yourself. The questions ask about your educational background and current work. Please answer all of the questions as completely as possible. Please remember your answers will remain confidential.

1a. What is your sex? _____
1 = Female
2 = Male

1b. What is your marital status? _____
1 = single 4 = separated
2 = married 5 = remarried
3 = divorced 6 = widowed

1c. How are you related to the study participant?

I am his/her: 1 = mother 6 = father
2 = stepmother 7 = stepfather
3 = grandmother 8 = grandfather
4 = aunt 9 = uncle
5 = female legal guardian 10 = male legal guardian
11 = other (please describe) _____

2. What is your date of birth? _____/_____/_____

3. Education: What is the highest level of education you have completed? (Please circle one)

1. Less than 7th grade
2. Junior high school (9th grade)
3. Partial high school (10th or 11th)
4. High school graduate
5. Trade school
6. Partial College (at least one year of specialized training)
7. College or university graduate (e.g. BA, BS)
8. Graduate of professional degree

4a. What kind of work are you doing? (What is your occupation?)

_____

(For example: Homemaker, retail sales, machinist, stock clerk, farmer, etc.)

4b. Is there another adult caregiver living in your home? (Please circle one)

YES NO **IF NO**, please go to #5.

4c. **IF YES**, what is their relationship to you and/or the study participant? (Please circle one)

(1) Husband/participant's biological father
(2) Wife/participant's biological mother
(3) Husband/participant's stepfather
(4) Wife/participant's stepmother
(5) My partner (boyfriend/girlfriend)
(6) Participant's grandmother
(7) Participant's grandfather
(8) Other: _____ (Please describe)

5. Approximately what is your present annual family income? (CIRCLE ONE)

1.	under $ 4,000	6.	$16,001--$20,000
2.	$ 4,000-- $ 7,000	7.	$20,001--$30,000
3.	$ 7,001-- $10,000	8.	$30,001--$50,000
4.	$10,001-- $13,000	9.	$50,001--$75,000
5.	$13,001-- $16,000	10.	Over $75,000
		11.	Prefer not to answer

6. Please list all of your children (starting with your child diagnosed with Sickle Cell Disease and any other children living in the home), and include birth date and relationship to your child. Also, please check if the child lives with you now.

RELATIONSHIP TO YOUR CHILD (sister, brother)	BIRTH DATE (mo/day/year)	Living with you now (Yes or No)
1. Participant Child		
2.		
3.		
4.		

7. Please list any other members of your family who <u>have Sickle Cell Disease</u> (including yourself, if appropriate).

RELATIONSHIP TO YOUR CHILD (parent, grandparent, cousin, etc)	SEX (male/ female)	Living with you now (Yes or No)	Has pain related to sickle cell disease? (Yes or No)
1.			
2.			
3.			
4.			

8. What is your race? _____

1 = White
2 = Black/African American
3 = Asian
4 = American Indian or Alaskan Native
5 = Native Hawaiian or other Pacific Islander
6 = Other: _____
7 = More than one race: _____
8 = Prefer not to answer

9. What is the race of your participant child? _____

1 = White
2 = Black/African American
3 = Asian
4 = American Indian or Alaskan Native
5 = Native Hawaiian or other Pacific Islander
6 = Other: _____
7 = More than one race: _____
8 = Prefer not to answer

MEDICAL HISTORY

1. What type of sickle cell disease does your child have?

   SS SB^0 thalassemia SB^+ thalassemia SC Other: _____ Not Sure

2. What treatments or medications is your child receiving for pain?

3. Approximately how many days has your child had Sickle Cell Disease pain **in the past month?** _____ day(s)

4. Approximately how many days has your child had Sickle Cell Disease pain **in the past year?** _____ day(s)

5. How many times has your child been to the Sickle Cell Clinic in the past year? ____

6. How many times has your child been to the Emergency Room in the past year? ____

7. How many times has your child been hospitalized (admitted) in the past year?

0 times	3 times	6 times
1 time	4 times	7 times
2 times	5 times	more than 7 times

8. How many pain crises (sickle cell pain lasting for at least 2 hours) has your child had in the last 12 months? _____

9. How many days of school did your child miss due to pain this past school year? ____ day(s)

10. How severe is your child's sickle cell disease?
    a) mild
    b) moderate
    c) severe

11. How much impact has your child's sickle cell disease had on your and your child's lives? (Circle appropriate number)

1	2	3	4	5
No Impact				Tremendous Impact

12. How would you rate your child's overall health? (Circle appropriate number)

1	2	3	4	5
Very Poor				Excellent

13. How confident are you that your child can effectively manage their illness on their own?

1	2	3	4	5
Not Confident At All				Very Confident

14. Have you or your child been involved in any other studies at Children's Healthcare of Atlanta?
    yes___ no___

15. Are you interested in participating in future studies at Children's Healthcare of Atlanta?
    yes___ no___

Appendix B. Facts About Your Pain (Child)

FACTS ABOUT YOUR PAIN (CHILD)

1. Please rate your pain by marking the box beside the number that best describes your pain at its **worst** in the last 2 weeks.

☐0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10
No pain Pain as Bad As You Can Imagine

2. Please rate your pain by marking the box beside the number that best describes your pain at its **least** in the last 2 weeks.

☐0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10
No pain Pain as Bad As You Can Imagine

3. Please rate your pain by marking the box beside the number that best describes your pain on the **average**.

☐0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10
No pain Pain as Bad As You Can Imagine

4. Please rate your pain by marking the box beside the number that tells how much pain you have **now**.

☐0 ☐ 1 ☐ 2 ☐ 3 ☐ 4 ☐ 5 ☐ 6 ☐ 7 ☐ 8 ☐ 9 ☐ 10
No pain Pain as Bad As You Can Imagine

5. How often do you have pain?
   - ☐ Everyday
   - ☐ 5-6 days per week
   - ☐ 3-4 days per week
   - ☐ 1-2 days per week
   - ☐ A few days per month
   - ☐ Other (explain): _____

6. How many days have you had pain in the past month? _____
   If you answered 15 or more days, please move on to #7.

7. If you had 15 or more days of pain in the past month, how long have you been having ongoing pain problems (pain on most days out of the month)?

   _____ months or _____ years

Appendix C. Pain Catastrophizing Scale – Child Version (PCS-C)

CHILD PAIN CATASTROPHIZING

Omitted in compliance with copyright legislation

Appendix D. Adult Responses to Children's Symptoms (ARCS)

PARENT RESPONSE TO CHILDREN'S PAIN SYMPTOMS

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Appendix E. Functional Disability Inventory – Child Form (FDI)

FUNCTIONAL DISABILITY

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Appendix F. Pediatric Quality of Life Sickle Cell Disease Module (Peds-QL SCD)

HEALTH-RELATED QUALITY OF LIFE

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