

Pain-related injustice appraisals in youth with sickle cell disease: a preliminary investigation

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3 Pain-related injustice appraisals in youth with sickle cell disease: a preliminary investigation
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Abstract

Objectives: Sickle cell disease (SCD) is a genetic disorder that affects approximately 100,000 Americans, the majority of whom are African American. SCD-related pain often has deleterious effects on functioning and quality of life. The inherited nature of SCD, SCD-related stigma, and serious physical and functional impact of SCD-related pain create a situation ripe for individuals to appraise their SCD-related pain as unfair or unjust. The aim of this preliminary investigation is to explore the extent to which pediatric patients with SCD appraise their pain as unjust and how these appraisals relate to functioning.

Methods: Participants were youth with SCD (N=30, mean age=11.3, 57% boys) who attended a hematology clinic visit. Patients were invited to complete paper-based questionnaires assessing pain-related injustice appraisals, pain catastrophizing, pain and hurt, functional disability, depression, anxiety, and peer relationships.

Results: Results of hierarchical regressions indicate that pain-related injustice significantly predicted functional disability, depression, and anxiety after controlling for patient pain and catastrophizing.

Conclusions: These findings suggest that pain-related injustice appraisals are an important contributor to the pain experience of youth with SCD. Early identification and remediation of pain-related injustice appraisals could have long-term functional benefits for youth with SCD.

Keywords: injustice, sickle cell, functioning, anxiety, depression, youth

Introduction

Sickle cell disease (SCD) is a genetic disorder that affects approximately 100,000 Americans, the majority of whom are African American[1]. SCD is a lifelong disease characterized by recurrent, unpredictable, and disabling pain due to vaso-occlusive crises (VOCs) [2,3] and progressive end-organ damage[4,5]. In addition to acute painful events, approximately 20% of children with SCD will suffer from chronic pain[6]. Combining painful events with additional sickle cell complications, the acute healthcare costs associated with hospital admissions for SCD patients exceeds \$2 billion/year [7] with an estimated \$900 million/year of that for treatment of pediatric patients with SCD in the U.S[8].

SCD-related acute and chronic pain have deleterious effects on functioning and quality of life for many children/adolescents with SCD[9,10,11,12,13,14,15]. Some youth with SCD require frequent hospitalizations and emergency department (ED) visits due to acute pain episodes[14]. Medical complications due to SCD often require numerous outpatient medical appointments with specialty services such as hematology, pulmonology, nephrology, and pain management[9]. In addition to these healthcare-related disruptions, acute and chronic pain in SCD interferes with functioning (cognitive and physical), sleep, mood, and quality of life[10,11,12,13]. Youth with SCD report worse physical functioning due to pain, particularly among youth reporting chronic, compared to episodic SCD-related pain[16]. Youth with SCD have been found to have a high incidence of mental health symptoms including anxiety and depression[17] which is associated with more frequent and longer hospitalizations for VOCs[18]. Children and adolescents with SCD face several barriers when it comes to academic success. SCD has detrimental effects on

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3 cognitive functioning, even without vascular complications (e.g., cerebral infarction)[19].

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5 Additionally, SCD-related acute and chronic pain interferes with school attendance, with an
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8 estimated 12-30 school days missed per year[20,21], which may significantly impact child
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10 cognitive and social functioning[22].
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16 The burden of SCD – pain-specific and more generally – falls on African Americans who
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18 already face a disproportionate number of injustices including poverty[23,24,25], racism
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20 [26,27,28,29], and biased criminal justice practices[30,31]. For youth with SCD, perceived
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22 racism is associated with higher levels of depressive symptoms and poorer health-related quality
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24 of life [32,33]. Studies have also identified that African Americans with SCD are at risk for
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26 suboptimal care[34], with healthcare providers' negative perceptions of patients (e.g. drug
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28 seeking) with SCD contributing to this risk. Opioid use and opioid-related hospitalizations have
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30 remained stable among SCD patients from 1998 to 2013. This is in contrast to the general
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32 population in the U.S. where opioid use and inpatient deaths from opioid-related admissions
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34 have risen over time[35]. Additionally, the prevalence of opioid abuse is lower in patients with
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36 SCD (0.5%-8%) than in other chronic pain conditions (3%-16%)[36]. Unfortunately, perceptions
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38 persist among providers that adolescent and adult patients with SCD are drug seeking and at
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40 higher risk for opioid abuse[34,37,38,39,40,41]. These false perceptions have been associated
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42 with longer wait times for analgesic administration and poorer provider adherence to the
43
44 National Institute of Health Lung and Blood Institute Guidelines for SCD treatment[42]. Youth
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46 and adult patients are aware of these provider perceptions and consequently feel mistrusted and
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48 stigmatized[43,44]. This is important because health-related stigma – defined as negative
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50 attribution or discrediting perception of a person or group based on their health status[45] – is
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3 associated with poorer health outcomes and quality of life in adults[46,47] and youth[48] with
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6 SCD.
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11 The inherited nature of SCD, the serious physical and functional impact of SCD-related pain, and
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13 the all too frequent stigma directed toward people with SCD may give rise to appraisals of
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15 unfairness or injustice (i.e., lack of fairness or justice). The focus of these appraisals can vary
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17 based on the lived experience of patients with SCD. For example, patients may perceive their
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19 (suboptimal) care as unjust. Ezenwa and colleagues[2] examined injustice appraisals surrounding
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21 healthcare provider treatment (i.e., healthcare injustice) and found that higher injustice appraisals
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23 were associated with more pain and stress in a sample of adults with SCD. Additionally, patients
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25 may regard their condition or symptoms, such as pain, as unfair or unjust. In the broader chronic
26
27 pain literature, pain-related injustice, or appraising one's pain as unfair or unjust, is associated
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29 with poorer physical[49,50,51,52,53,54], emotional[53,55,56] and occupational
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31 functioning[52,57,58] in adults with various pain conditions. Similar relationships have found in
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33 children and adolescents suffering from chronic pain[59,60].
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43 Despite its seeming relevance, pain-related injustice has yet to be examined in youth with SCD.

44 This is surprising given that the stigma[33,48] and negative outcomes[10,11,12,13] experienced
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46 by youth with SCD create a situation ripe for injustice appraisals. Given the severity of medical
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48 and psychosocial complications of SCD and that the majority of patients with SCD experience
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50 pain, it is critically important to identify appraisals, such as pain-related injustice, that contribute
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52 to worse functional outcomes for individuals, particularly youth with SCD. The aims of this
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3 study are to: 1) assess the extent to which pediatric patients with SCD appraise their pain as
4 unjust, 2) compare levels of injustice in pediatric patients with SCD to other samples of youth
5 with non-SCD chronic pain, and 3) investigate how these injustice appraisals uniquely relate to
6 functioning (functional disability, depression, anxiety, and peer relationships) in pediatric
7 patients with SCD. We hypothesize that pain-related injustice will be related to higher levels of
8 disability, depression, and anxiety, as well as worse peer relationships, among children and
9 adolescents with SCD.
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24 **Materials and Methods**

25 **Participants**

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29 Participants were children and adolescents along with their caregiver who attended a scheduled
30 hematology clinic visit at Children's Hospital of Alabama during 2018 and 2019. Potentially
31 eligible youths were identified through medical chart review. Eligible participants and their
32 caregivers were approached by research staff during regular hematology clinic visits while
33 waiting to see the hematologist for SCD-related care. Approximately 5 families declined to
34 participate. All individuals provided consent and/or assent (if youth was younger than 14 years
35 old) if they wanted to participate. All individuals who consented completed the study. The
36 sample included 30 children with SCD and their caregiver. Inclusion criteria were: (a) diagnosis
37 of SCD; (b) child age between 8-17 years old; (c) no known history of overt stroke (via medical
38 record review); and (d) have a Full Scale IQ over 70. Exclusion criteria were children and/or
39 parents who did not speak English.
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Procedure

All eligible patients and caregivers who expressed interest in completing the questionnaires provided written assent and informed consent prior to study participation. Patients and their caregivers completed paper-based questionnaires in the SCD clinic. Individuals were compensated with \$25 cash for each completed packet (\$25 for the parent packet and \$25 for child packet) at the completion of the study. The Institutional Review Board of the University of Alabama at Birmingham approved the study.

Measures

Socio-demographic Characteristics

Socio-demographic data including genotype, age, sex, race/ethnicity, and current grade were collected from the child's caregiver.

Injustice Experiences Questionnaire

Pain-related injustice appraisals were assessed using the Injustice Experiences Questionnaire[50]. The IEQ is a 12-item measure that assesses appraisals of injustice related to one's own pain experience. Using a 5-point Likert scale ranging from 0 ("never") to 4 ("all the time"), child participants rate how frequently they identify with statements such as "I should not have to live this way." Item values are summed to calculate a total score. The instructions for the original measure directed respondents to focus on "thoughts and feelings you may experience when you think about your injury." For this study, the instructions from the original measure

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3 were modified so that child participants were directed to respond to items in reference to their
4
5 own pain. This measure has been used in youth with mixed chronic pain conditions and
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7 demonstrated similar factor structure to the original measure[59] and good reliability[59,60]. The
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9 measure demonstrated good internal consistency in the current sample ($\alpha=.92$).
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16 *Pain Catastrophizing Scale – Child Version*

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19 The Pain Catastrophizing Scale for Children (PCS-C) was used to assess pain
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21 catastrophizing[61]. The PCS-C is a 13-item questionnaire adapted from the Pain
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23 Catastrophizing Scale (PCS)[62]. The PCS-C assesses the degree an individual engages in
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25 rumination, magnification, and feelings of helplessness when in pain. Individuals rate statements
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27 on a 5-point Likert scale ranging from 0 (“not at all”) to 4 (“extremely”). Responses are summed
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29 to calculate a total score. The PCS-C has previously been used in sickle cell samples [63] and
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31 showed good internal consistency in the current sample ($\alpha=.90$).
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40 *Pediatric Quality of Life Inventory – Sickle Cell Module – Pain and Hurt Subscale*

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42 Patients' pain was assessed using the Pediatric Quality of Life Inventory Sickle Cell Disease
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44 Module (PedsQL-SCD)[64,65] Pain and Hurt subscale. The subscale includes 9 self-report items
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46 about pain and hurt (e.g., “I hurt a lot” and “I have pain everyday”), which are rated using a 5-
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48 point Likert scale ranging from 0 (“never”) to 4 (“almost always”). Items are reverse scored and
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50 transformed into a 0 to 100 scale. The PedsQL-SCD Pain and Hurt subscale score is calculated
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52 by averaging the 9-items, with higher scores reflecting less pain in the individual’s life. The Pain
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3 and Hurt subscale score and clinical classifications (81 – 100 = high levels of HRQOL, 61 – 80 =
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5 intermediate levels HRQOL, and 0 – 60 = poor HRQOL related to pain) were used in the current
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7 analyses[66]. Reliability and validity of the Pain and Hurt subscale score have been previously
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9 established [64]. The PedsQL-SCD Pain and Hurt subscale has previously been used in sickle
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11 cell samples [64,65,66] and demonstrated adequate internal consistency in the current sample
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15 ($\alpha=.80$).

21 *Outcome Variables*

24 *Functional Disability Inventory*

27 The level of functional limitation for each patient was measured using the Functional
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29 Disability Inventory (FDI)[67]. The FDI is an established and commonly used tool for children
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31 and adolescents who have chronic pain[68]. Fifteen items assessing difficulty in performing
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33 daily tasks, such as being at school all day and walking to the bathroom, are rated on a scale of 0
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35 (“no trouble at all”) to 4 (“impossible”). The items are summed to create a total score, with
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37 higher scores reflecting greater functional disability. The FDI has previously been used in sickle
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39 cell samples [69,70] and had good internal consistency in the current sample ($\alpha=.86$).

47 *PROMIS Measures (depression, anxiety, and peer relationships)*

50 PROMIS pediatric measures have been developed using qualitative and item response theory
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52 methods and validated in children with SCD[71,72,73]. Validated short forms of the depressive
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54 symptoms, anxiety, and peer relationships[74] scales were administered in this study. These
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3 measures assess a child's report of their depressive (e.g., "I could not stop feeling sad") and
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5 anxiety (e.g., "I felt like something awful might happen") symptoms, as well as social
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7 functioning (e.g., "other kids wanted to be my friend") over the previous seven days. All scale
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9 items included in this study used standardized five-point response options (never, almost never,
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11 sometimes, often, almost always). Items on each form are summed with higher scores indicating
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13 more of what is being measured. For depression and anxiety, this means higher scores indicate
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15 worse depression or anxiety. For peer relationships, this means higher scores indicate better peer
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17 relationships. Raw scores were transformed into standard scores for analyses. PROMIS measures
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19 have previously been used in sickle cell samples [71,72]. All PROMIS measures demonstrated
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21 good internal consistency (Depression: $\alpha=.96$; Anxiety: $\alpha=.86$; Peer Relationships: $\alpha=.85$)
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30 **Statistical Analyses**

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33 A summary independent samples t-test was used to compare pain-related IEQ scores between the
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35 current and a previously reported pediatric sample of children with chronic pain[60]. Zero-order
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37 correlations were calculated to assess the bivariate relationship between perceived injustice, pain
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39 catastrophizing, and outcomes variables (functional disability, depression, anxiety, and peer
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41 relationships). Hierarchical regressions were used to examine the multivariate relationship
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43 between perceived injustice and outcomes variables including functional disability, depression,
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45 anxiety, and peer relationships). The contribution of pain and hurt (step 1), and pain
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47 catastrophizing (step 2) were added as covariates so that the unique role of injustice appraisals
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49 can be determined (step 3). Previous literature has identified pain catastrophizing and pain-
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51 related injustice are highly correlated[50]. In preliminary analyses, age and gender were explored
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3 as covariates and were not found to be significant predictors in the models ($p>.05$). A power
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5 analysis specified that 27 participants were required to detect a medium effect ($d=.50$) at 80%
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7 power with three predictors in the regression[75]. Therefore age and gender were removed as
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9 covariates in favor of adequately powered statistical analyses. The variance-inflation factors
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11 (VIF) of all regression analyses were acceptable (range 1.31–2.54), suggesting that
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13 multicollinearity was not an issue. Standardized beta weights were assessed to determine the
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15 strongest predictor in each regression. All analyses were run using IBM SPSS Version 26[76].
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23 **Results**

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26 Sample descriptives are summarized in Table 1. The final sample included 30 children and their
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28 caregivers; there were slightly more males ($N=17$) than females ($N=13$), and all participants
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30 identified as Non-Hispanic African American. Majority of caregivers identified as the maternal
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32 caregiver ($N=26$). Average age of the sample was approximately 11.3 years ($SD=2.73$).
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36 Descriptive information for measures is detailed in Table 2. Using established clinical cutoffs, on
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38 average the sample reported a moderate level of catastrophizing[77], and intermediate/moderate
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40 level of pain and hurt[66]. In line with other SCD samples [69,70], 63% of the sample reported
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42 minimal, 30% reported moderate, and 7% reported a high degree of functional disability
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44 according to established clinical cutoffs [68]. The sample average was not clinically elevated on
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46 the PROMIS depression, anxiety, or peer relationships scales, which is in line with previous
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48 investigations in SCD samples [71,73]. However, a portion of the sample reported elevated
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50 scores on the PROMIS depression (30%), anxiety (30%), and peer relationships (36%) scales.
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54 Pain-related injustice did not differ based on patient sex ($p>.05$). When compared to a sample of
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3 youth with mixed chronic pain conditions (75% female, 83% White, $M_{age}=14.1$ years,
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5 $SD=2.25$)[60], the current SCD sample reported significantly lower injustice appraisals
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7 ($t(195)=8.46, p<.01, d=.67$). This difference may be related to differences on several
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9 demographic variables between the two samples (e.g., gender, race, age). However, because
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11 systematic differences in pain-related injustice across gender, race, and age have yet to be
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13 established in the literature, this finding (lower injustice appraisals among the current sample of
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15 patients with SCD) should be interpreted with caution.
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23 *Bivariate relationships between pain-related injustice and pain-related factors/outcomes*

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26 The results of bivariate correlation analyses are presented in Table 2. Higher levels of pain-
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28 related injustice were significantly associated with higher levels of pain catastrophizing ($r=.58$),
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30 functional disability ($r=.70$), depression ($r=.60$), and anxiety ($r=.72$), as well as worse pain ($r=-$
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32 $.44$). Pain-related injustice was not significantly related to age or peer relationships ($ps>.05$).
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45 *Hierarchical regressions explaining functional disability, depression, anxiety, and peer* 46 47 *relationships*

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49 **Functional disability.** In step 1 of the analysis, pain and hurt did not account for a significant
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51 amount of the variance in functional disability ($F(1, 28)=3.73, p=.06$, Table 3). In step 2, the
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53 addition of catastrophizing was significant, accounting for 18% of unique variance in functional
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55 disability ($F(2,27)=5.80, p<0.01$, Table 3). In step 3, the addition of pain-related injustice
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57 accounted for an additional 21% of the variance in functional disability, above and beyond that
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3 accounted for by pain and hurt and catastrophizing ($F(3,26)=8.97, p<.01$, Table 3). Pain-related
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5 injustice was the only significant predictor in the final model.
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11 **Depression.** Pain and hurt ($F(1, 28)=3.94, p=.06$, Table 3) did not account for significant
12 variance in depression. In step 2, the addition of catastrophizing resulted in a significant model
13 ($F(2, 27)=5.69, p<.01$, Table 3) and accounted for an additional 17% of unique variance in
14 depression. In step 3 of the model, pain-related injustice accounted for an additional 12% of the
15 variance in depression ($F(2, 26)=6.07, p<.01$, Table 3). Pain-related injustice was the only
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Anxiety. In step 1, pain and hurt did not account for a significant amount of the variance in
anxiety ($F(1, 28)=2.60, p=.12$, Table 3). In step 2, catastrophizing accounted for an additional
30% of unique variance in anxiety ($F(2, 27)=8.60, p<.01$, Table 3). The addition of pain-related
injustice in step 4 accounted for an additional 20% of the variance in anxiety above and beyond
that accounted for by pain and hurt and catastrophizing ($F(3, 26)=12.34, p<.001$, Table 3). Pain-
related injustice and catastrophizing were both significant predictors in the final model.

Peer relationships. Pain and hurt, catastrophizing, and pain-related injustice were not
significantly associated with peer relationships ($ps>.05$, Table 3).

Discussion

The results of this preliminary study of youth with SCD indicated that higher pain-related injustice was related to worse pain and greater pain catastrophizing, functional disability, depression, and anxiety. Additionally, after controlling for pain and catastrophizing, pain-related injustice appraisals contributed to worse physical functioning and symptoms of anxiety and depression. By contrast, quality of peer relationships was not significantly associated with pain-related injustice appraisals. Collectively, these preliminary findings suggest that pain-related injustice is a clinically meaningful contributor to the pain experience of youth with SCD.

Pain-related injustice demonstrated the strongest relationship with functional disability, aligning with previous studies in adults and children[50,51,59,60]. This relationship was considerably stronger than that observed in previous investigations of children with mixed chronic pain[59,60], which may indicate that pain-related injustice is more closely tied to physical functioning for patients with SCD. Pain behaviors (e.g., guarding, bracing) may be one pathway through which pain-related injustice is related to poor physical functioning. Pain behaviors have been associated with increased pain-related injustice[78,79] and have been identified as a mediator in the relationship between pain-related injustice and disability in adults with chronic pain[51]. For individuals who endorse IEQ items such as “most people don’t understand how serious my condition is,” pain behaviors may serve to communicate to others how severe their pain is. However, such pain behaviors may have functional consequences, as they associated with greater disability in children and adolescents[80]. To date, no study has investigated if the connection between injustice, pain behaviors, and physical disability replicates in youth with

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2
3 chronic pain. Additionally, the degree of social support received by youth with SCD may buffer
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5 the relationship between pain-related injustice and functional disability. In a sample of adults
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7 with chronic pain and HIV, a significant association between injustice and pain interference was
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9 supported only for individuals endorsing low level of social support[81]. This is particularly
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11 important for children and adolescents who rely heavily on parents and other close family
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13 members to access proper SCD care.
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21 Similar to previous investigations of emotional distress in adult [50,78,82] and youth samples
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23 [59,60], pain-related injustice significantly predicted anxiety and depression. VOCs are
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25 unpredictable and may leave children worrying about when the next crisis will occur[83]. Other
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27 serious SCD-related events such as a stroke or acute chest syndrome, or socially uncomfortable
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29 SCD-related symptoms such as priapism[4] may also cause youth to ruminate and worry about
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31 the occurrence of these events and symptoms. Moreover, previous studies in youth with SCD
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33 found that negative thinking (i.e., catastrophizing, fear, anger) compromises one's ability to cope
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35 with pain and is related to poorer adjustment to painful episodes[84,85,86], as well as to
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37 increased anxiety and depressive symptoms[84]. Pain-related injustice appraisals represent
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39 another form of negative thinking and, thus, may similarly exacerbate anxiety and depressive
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41 symptoms. For example, an adolescent who endorses the belief "most people don't understand
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43 how severe my condition is" (Item 1 on IEQ) may experience greater worry and anxiety that
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45 proximal others will not take their pain seriously or help them during a VOC. Similarly, thoughts
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47 such as "nothing will ever make up for all that I have gone through" (Item 9 on IEQ) may
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49 engender feelings of hopelessness and sadness, both facets of depression[87,88,89]
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6 Pain-related injustice was not significantly associated with peer relationships. This diverges from
7 previous findings in a sample of mixed pain conditions [59,60]. Although these results should be
8 interpreted with caution, several key differences between a sickle cell diagnosis and other
9 chronic pediatric pain conditions may explain this divergence. A diagnosis of sickle cell is
10 confirmed via blood test at birth[90]. Diagnosis confirmation, particularly through objective
11 tests, could legitimize an individual's condition and may result in less disbelief or invalidation
12 from close friends or family members[91,92]. This feature of SCD stands in contrast to many
13 other pain conditions that lack diagnostic certainty[92] and, thus, elicit skepticism and dismissal
14 about their diagnosis from others[93]. Additionally, given SCD is a well-known racially-
15 weighted disease, same-race peers may have greater empathy for youth affected by SCD.
16 Previous studies have documented that observers demonstrate greater empathy for same-race
17 compared to other-race individuals in pain[94,95]. Thus, peers may have greater empathy for
18 youth affected by SCD and this may influence the way peers interact with patients with SCD
19 compared to other less-known pediatric pain conditions. However, important to note, SCD can
20 have apparent physical (e.g., short stature) and cognitive signs that might put youth with SCD at
21 risk for teasing or bullying[96].
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47 Of note, the level of pain-related injustice endorsed in the current sample was significantly lower
48 than that of previous samples of youth with mixed chronic pain conditions [59,60]. Several
49 differences between the samples may explain this result. The current sample is younger (*M*
50 *age*=11.3) than previous samples (*M age*=15 & 14, respectively) which bears heavily both on
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3 current developmental stage (e.g., adolescence) and typical life experiences. As adolescence is a
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5 time of increased independence, pain may increasingly impede age-related milestones (i.e.
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7 development of friendships, educational advancement, and independence from parents),
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9 increasing the likelihood that pain is appraised as unjust. Alternatively, the episodic nature of
10
11 SCD-related pain, may result in weaker injustice appraisals compared to conditions that are
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13 characterized by more consistent pain(e.g., Juvenile Idiopathic Arthritis, Complex Regional Pain
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15 Syndrome). However, as the likelihood of chronic SCD-related pain increases with age, so too
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17 may injustice appraisals.
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26 Although these results are preliminary and should be replicated in a larger sample, taken together
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28 with previous studies [59,60], they indicate that pain-related injustice may be an important
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30 component of the chronic pain experience for many youth. Sickle cell disease is a lifelong
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32 condition. Therefore, early identification and remediation of pain-related injustice appraisals
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34 could have long-term functional benefits for youth with SCD. Psychologists have a vital role in
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36 helping patients address these negative appraisals. An initial step would be examining whether
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38 existing psychotherapy interventions, such as Cognitive Behavioral Therapy (CBT) [97] and
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40 Acceptance and Commitment Therapy (ACT) [98], can target pain-related injustice appraisals,
41
42 similar to other negative appraisals (e.g., pain catastrophizing). Additionally, outside of the
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44 therapy context, strategies to reduce unjust experiences in acute care settings during VOCs have
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46 been proposed. One strategy, being used at some institutions, is the creation of individualized
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48 VOC pain protocols as a way to standardize treatment, promote partnership between healthcare
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50 providers and patients, and reduce opportunities for invalidating and unjust experiences[41].
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55 Another strategy is to develop educational modules – focused on common situations or provider-

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3 patient interactions that result in appraisals of injustice – for healthcare trainees and providers
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5 [99] so that they may gain insight into how their actions drive injustice appraisals and
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7 consequently worse outcomes for patients with SCD.
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14 The current study is not without limitations. The small sample size reduces the power to detect
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16 small but significant associations between study variables. However, despite the sample size,
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18 pain-related injustice appraisals significantly predicted physical functioning and anxiety, even
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20 after controlling for pain catastrophizing. The strong relationships identified between pain-
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22 related injustice and important functional outcomes, as well as the previously established
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24 reliability[59,60] and validity[59] of the measure in samples of youth with pain point to its utility
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26 in measuring pain-related injustice. However, the current IEQ was adapted from a measure
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28 created for adults with pain due to injury. Therefore, the IEQ may not capture all facets of what
29
30 youth appraise as unjust about their pain experiences. Another limitation is that other factors
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32 such as demographic factors, SCD genotype, healthcare utilization, and socioeconomic status
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34 were not be explored in the current study. Relatedly, SCD occurs within the context of many
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36 racially-related injustices (i.e., poverty, discrimination, biased criminal justice practices), which
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38 were not assessed in this study but may contribute to the development or maintenance of pain-
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40 related injustice appraisals and/or contribute to poor functional and psychosocial outcomes. A
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42 recent study in a sample of adults with chronic low back pain found that pain-related injustice
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44 mediated the relationships between past discriminatory experiences and disability, as well as past
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46 discriminatory experiences and depression for African American participants[100]. Lastly, all
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48 study participants were from a single center in the South. Therefore, findings may not generalize
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50 to other youth affected by SCD.
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7 As our study was a preliminary step in examining the relationships between pain-related injustice
8 appraisals, pain, and functional outcomes in youth with SCD, numerous future directions remain.
9 Possible precursors to pain-related injustice appraisals, including perceptions of racial bias and
10 health-related stigma surrounding a sickle cell diagnosis, should be examined. The existing
11 literature suggests both are common and contribute to poorer quality of life and health outcomes
12 in youth with SCD[32,33,48]. Parental pain-related injustice appraisals about their child's pain
13 should also be examined. Previous research has found discordant parent-child injustice appraisals
14 are associated with worse functional outcomes, particularly when the child or adolescent
15 perceives their pain as unjust and the parent does not[60]. Due to the genetic nature of SCD, a
16 youth with SCD may have a parent or family member with SCD. This parallel disease experience
17 may foster shared appraisals of injustice about SCD. Additionally, youth may mirror coping
18 strategies demonstrated by family members with SCD-related pain. As mentioned above, African
19 Americans face compounding injustices. The impact that discriminatory experiences, both
20 general and health-care related, have on SCD trajectory and patient functioning, and the
21 relationship with pain-related injustice should be assessed. Lastly, the differences between
22 findings in the current study and previous pain-related injustice investigations in pediatric mixed
23 chronic pain samples highlight the importance of examining relationships in condition specific
24 samples. Due to differences (e.g., pain severity and fluctuation, required medical care, functional
25 limitations) across pain conditions, the strength of relationships between pain-related injustice
26 and functional outcomes may vary.
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5 The authors have nothing to disclose. We confirm that there have been no closely related
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Table 1. Demographic Characteristics

N=30		n (%)
Sex		
	Male	17 (56.7)
	Female	13 (43.3)
Race		
	Non-Hispanic African-American	30 (100)
Age		
	8-10 years	13 (43.3)
	11-13 years	9 (30.0)
	14-16 years	8 (26.7)
Genotype		
	HbSS	63%
	HbSC	23%
	HbSB+ Thalassemia	10%
	Not reported	1%

Table 2. Zero-order correlations among study variables.

	Mean	SD	1	2	3	4	5	6
1. Injustice Experience Questionnaire - Child	11.17	12.42	1					
2. PedsQL - Sickle Cell Pain and Hurt Subscale	68.43	19.11	-0.44*	1				
3. Pain Catastrophizing Scale - Child Report	20.13	13.3	0.58**	-0.41*	1			
4. Functional Disability Inventory	11.10	10.73	0.70**	0.34	0.53**	1		
5. PROMIS Depression (t-scores)	45.63	13.33	0.60**	-0.35	0.52**	0.40*	1	
6. PROMIS Anxiety (t-scores)	43.49	11.22	.072**	-0.29	0.62**	0.73**	0.69**	1
7. PROMIS Peer Relationships (t-scores)	44.78	9.47	-0.08	-0.10	-0.15	-0.06	-0.23	0.07

Abbreviations: PedsQL, Pediatric Quality of Life Inventory; PROMIS, Patient-Reported Outcomes Measurement Information System

* $p < 0.05$, ** $p < 0.01$ * $p < 0.05$, ** $p < 0.01$

Table 3. Regression models

		B	β	SE	t	R²	Δ R²
Regression 1: Dependent = Functional Disability							
Step 1						0.09	-
	Pain and Hurt	-0.19	-0.34	0.10	-1.93		
Step 2						0.30**	0.18*
	Pain and Hurt	-0.08	-0.15	0.10	-0.85		
	Catastrophizing	0.38	0.47	0.14	2.66*		
Step 3						0.51**	0.21**
	Pain and Hurt	-0.01	-0.01	0.09	-0.72		
	Catastrophizing	0.16	0.19	0.14	1.12		
	Pain-related Injustice	0.50	0.58	0.15	3.32**		
Regression 2: Dependent = Depression							
Step 1						0.12	-
	Pain and Hurt	-0.25	-0.35	0.12	-1.98		
Step 2						0.30**	0.17*
	Pain and Hurt	-0.11	-0.16	0.12	-0.92		
	Catastrophizing	0.46	0.46	0.18	2.58*		
Step 3						0.41**	0.12*
	Pain and Hurt	-0.04	-0.06	0.12	-0.35		
	Catastrophizing	0.25	0.25	0.19	1.33		
	Pain-related Injustice	0.46	0.43	0.21	2.26*		
Regression 3: Dependent = Anxiety							
Step 1						0.09	-
	Pain and Hurt	-0.17	-0.29	0.11	-1.61		
Step 2						0.39**	.30**
	Pain and Hurt	-0.03	-0.04	0.1	-0.26		
	Catastrophizing	0.51	0.61	0.14	3.67**		
Step 3						0.59**	0.20**
	Pain and Hurt	0.05	0.09	0.08	0.65		
	Catastrophizing	0.28	0.34	0.13	2.13*		
	Pain-related Injustice	0.51	0.57	0.14	3.54**		
Regression 4: Dependent = Peer Relationships							
Step 1						0.01	-
	Pain and Hurt	0.05	0.10	0.10	0.53		
Step 2						0.02	0.01
	Pain and Hurt	0.03	0.05	0.11	0.23		
	Catastrophizing	-0.09	-0.12	0.15	-0.58		
Step 3						0.02	0.00
	Pain and Hurt	0.03	0.05	0.11	0.24		
	Catastrophizing	-0.10	-0.14	0.18	-0.54		
	Pain-related Injustice	0.02	0.02	0.19	0.09		

* $p < 0.05$, ** $p < 0.01$