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Emotion-Focused Avoidance Coping Mediates the Association between Pain and Health-Related Quality of Life in Children with Sickle Cell Disease

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Abstract

Sickle cell disease (SCD) is associated with pain and decreased health-related quality of life (HRQOL). Coping strategies influence pain but have not been evaluated as mediating the relation between pain and HRQOL in pediatric SCD. The current study examined whether pain-related coping mediates the association between pain and HRQOL in children and adolescents with SCD. 104 children and adolescents ages 8 to 18 years old ($M_{age}=12.93$ years) with SCD attending outpatient clinics completed pain intensity, HRQOL, and pain-related coping measures. Multiple mediation analyses were used to examine whether pain-related coping mediated the pain and HRQOL relation and whether types of coping (i.e., Approach, Emotion-Focused Avoidance, Problem-Focused Avoidance) were independent mediators. Total indirect effects for models examining Physical and Psychosocial HRQOL were not significant. After controlling for covariates, Emotion-Focused Avoidance significantly mediated the association between pain and Physical HRQOL (Effect=-.023, Bootstrapped SE=.018, 95% CI=-.0751,-.0003) but not the pain and Psychosocial HRQOL relation. Approach and Problem-Focused Avoidance were not significant mediators. Coping with pain in pediatric SCD is an important avenue for clinical intervention and additional research. Among children with SCD reporting high pain intensity, interventions should emphasize negative impacts of Emotion-Focused Avoidance coping and integrate other empirically supported coping strategies to improve HRQOL.

Keywords

Sickle Cell Disease; Coping; Health-Related Quality of Life; Children

Introduction

Sickle cell disease (SCD) is a group of genetic hematological conditions that affects approximately 1 in 500 African Americans,¹ making it one of the most common genetic diseases in the United States.² About 1,000 infants born in the United States are diagnosed with SCD each year.³ SCD is a progressive disease that affects multiple organ systems. One of the hallmark and debilitating characteristics of SCD is vaso-occlusive pain that result when sickled blood cells collect and inhibit blood flow.⁴ While specific triggers of these acute pain crises have yet to be identified, some research suggests that environmental and psychological factors such as cold temperatures, low barometric pressure, stress, and fatigue may play a role.^{2,5} In addition to acute pain, patients with SCD may also experience chronic pain as a result of musculoskeletal complications.⁴ In children, acute and chronic pain negatively impact functioning in a variety of social and emotional areas.^{6,7} As persons with SCD age and pain persists, a cycle of pain, inadequate coping skills, and disrupted functioning may develop.⁸ There is a need for more research to improve our understanding of pain and functioning in youth with SCD to identify children who may be at risk for poor health-related quality of life (HRQOL) outcomes and to inform the development of interventions to improve health and functioning.

HRQOL is considered a multidimensional patient centered construct⁹ and is recognized as an important aspect of functioning in children with chronic medical conditions. Poor HRQOL is associated with increased disease severity, behavioral and emotional problems, and higher healthcare utilization in children with a variety of medical conditions.^{10,11} Children with SCD have significantly lower HRQOL compared to healthy African American children in physical, psychological, and social domains of functioning.¹² When compared to children with cancer, hospitalized children with SCD have significantly lower total HRQOL, as well as in social- and self-competence domains, but higher emotional stability.¹³ In adults and children with SCD, pain significantly impacts multiple dimensions of HRQOL.^{14,15} In adolescents with SCD, pain negatively predicts aspects of HRQOL and disease-related parental stress has been found to mediate the relationship between pain and HRQOL.¹⁶ However, child internalizing symptoms did not mediate the relationship between pain and HRQOL in this study; thus, there remains an important need to examine other child factors, such as coping skills, that may mediate the association between pain and HRQOL in children with SCD. Coping skills in particular can be an important target for intervention; thus, better understanding of its role is critical.

Pain-related coping may be an important factor to consider to further understand the relationship between pain and HRQOL in children with SCD. Coping is defined as cognitive and behavioral efforts to manage external and internal demands that are stressful.¹⁷ Coping is considered a dynamic process that may change depending on characteristics of the stressor.¹⁸ Others have expanded on this definition by describing coping as conscious,

purposeful efforts to regulate emotion, cognition, behavior, physiology, and environment in response to stress.¹⁹ Different dimensions of coping have been identified, resulting in different types of coping styles. Coping consists of strategies that are problem-focused (i.e., making efforts to change some aspect of a situation by eliminating or altering it) and/or emotion-focused (i.e., strategies used to regulate the negative emotions associated with a situation).¹⁷ Another common dimension of coping is approach and avoidance.¹⁹ Approach coping refers to efforts to focus on or confront a stressful event (e.g., use techniques to minimize pain during a vaso-occlusive episode), whereas avoidance coping refers to attempts to avoid or ignore a stressful event.^{20,21} Generally, problem-focused and approach coping are associated with better health and adjustment outcomes, whereas emotion-focused and avoidance strategies relate to poorer outcomes.¹⁹ Coping has been linked to health status and is viewed as a central factor related to the well-being of children with chronic illnesses.²² The risk and resistance model of adjustment is a theory that has been applied to pediatric chronic medical conditions, including SCD. The theory posits that adjustment to a chronic medical condition is determined by risk factors such as disease severity and complications, daily living skills, and psychosocial stressors, as well as protective factors, such as intrapersonal/social support, social-ecological factors (e.g., family functioning, economic status), and stress processing (e.g., locus of control, coping strategies).²³ Thus, coping can be considered both a risk and resistance factor in predicting adjustment to pediatric medical conditions.⁴ Examining coping in children with SCD is important because how children cope with pain may influence their everyday psychosocial functioning and adjustment to SCD as adults.²⁴ In addition, interventions might be developed and implemented to modify children's pain-related coping to enhance adjustment.

Among children and adolescents with SCD, the use of mostly avoidant or passive pain coping strategies, such as resting and drinking fluids, is associated with more emergency room visits, lower activity levels, and higher pain intensity, whereas the use of active or approach coping strategies, such as distraction and increased activity, is associated with fewer emergency room visits and higher activity levels.^{24,25} In addition, children and adolescents with high levels of negative thinking, such as fear and anxiety, and those who engage in avoidance coping are less active and have poorer psychological adjustment.^{24,26,27} Coping has been examined as a mediator in the relationship between pain and psychological functioning in adolescents with SCD. Barakat et al. (2008) examined whether coping mediated the association between pain and internalizing symptoms. They found that negative thinking mediated the association between pain and depressive symptoms. However, negative thinking did not consistently mediate the association between pain and anxiety symptoms. Other dimensions of adjustment, such as physical functioning or HRQOL were not examined.

To date, only one research study has examined pain, coping, and HRQOL in children with SCD. Fuggle et al. (1996) found that SCD related pain impaired daily activities in children 6 to 16 years of age when compared to healthy control children.¹⁵ In addition, the most common pain coping strategy involved talking with parents about the pain and taking medication. However, this study did not utilize a multidimensional assessment of quality of life and coping was assessed utilizing diaries that did not assess specific types and dimensions of coping (approach, problem-focused, etc.) and associations between the

constructs were not examined. Thus, the purpose of the current study was to examine associations between pain, pain-related coping, and HRQOL in children with SCD. Specifically, it was hypothesized that (1) child report of pain would be associated with HRQOL, (2) approach coping would be associated with higher HRQOL and avoidance coping would be associated with lower HRQOL, and (3) coping style would mediate the relationship between child-reported pain and HRQOL.

Materials and Methods

Participants

Participants included 104 children diagnosed with SCD who ranged in age from 8 to 18 years of age ($M = 12.93$ years, $SD = 3.17$ years) who were receiving SCD-related care at two children's medical facilities and their caregivers. Both clinics were located in an urban city in the southeastern United States.

115 children and their families were approached to participate. Nine families (7.8%) did not wish to participate. Reasons for non-participation included not being interested in participating in research (7) and not having enough time to complete study measures (2). Therefore, 106 participants were enrolled in the study. However, 2 children (1.9%) did not complete the main outcome measures due to time constraints and were removed from analyses. Therefore, the final sample consisted of 104 children with SCD and their parents (90.4% enrollment rate).

Procedures

Children scheduled to receive routine SCD-related outpatient medical care at two urban children's hospitals and their parents were sequentially informed of the study by clinic personnel and directed to receive additional information from a trained research staff member who further explained the study and obtained parent consent and child assent if the family was interested in participating. Before meeting with physicians, the child and caregiver completed measures in a quiet room in the clinic. Children completed questionnaires regarding pain, pain-related coping, and HRQOL. Parents completed a demographics questionnaire and at the end of the medical visit, a nurse provided a rating of the child's disease severity. The governing IRBs approved this study.

Measures

Pain.—The Pediatric Pain Questionnaire (PPQ) is a self-report questionnaire used to assess pain from the child's perspective.²⁸ The PPQ consists of visual analogue scales (VAS) and open-ended questions. The VASs included on the PPQ are 100 mm horizontal lines that measure present pain and worst pain in the past week. The VAS questions are anchored at each end of the line with developmentally appropriate pain descriptions (e.g., No pain) and happy and sad faces. Scores on the PPQ range from 0 to 100, with higher scores representing more pain. The PPQ has demonstrated adequate reliability and validity in children with juvenile rheumatoid arthritis²⁸ and demonstrated validity in a sample of children with SCD and their parents.²⁹ The PPQ is considered a "well-established" instrument for assessing children's pain.³⁰ The Worst Pain (in the past week) VAS item has been used in previous

pediatric pain research³¹ and in preliminary analyses was more strongly associated with physical HRQOL ($r = -.43, p < .001$) compared to present pain intensity ($r = -.40, p < .001$). Thus, worst pain was utilized as the primary independent variable in study analyses.

Health-Related Quality of Life.—The Pediatric Quality of Life Inventory (PedsQL) 4.0 Generic Core was completed by children to assess HRQOL.³² The PedsQL is a 23-item measure that assesses several domains of functioning (e.g., Physical, Emotional, Social, and School Scales) and utilizes a 5-point Likert scale (0 = *never* to 4 = *almost always*). Scores on the PedsQL are reverse scored and transformed to scores ranging from 0 to 100, with higher scores representing better HRQOL. In addition to specific scaled scores, the items are averaged to create a Psychosocial Scale (i.e., Emotional, Social, and School Scales). Child-report on the PedsQL is valid and reliable (Cronbach's alphas ranged from 0.68 to 0.88)³² and the measure is considered "well-established".³³ In this sample of children with SCD, Cronbach's alpha for the PedsQL Physical Scale was 0.86 and 0.85 for the Psychosocial Scale and both of these scales were utilized in analyses.

Pain-related Coping.—To assess children's pain-related coping strategies, the Pain Coping Questionnaire (PCQ) was utilized.³⁴ The PCQ is a 39-item measure created for use with children 8 to 18 years of age. Children answered questions based on how often they engage in a particular coping strategy by using a 5-point likert scale (1 = *never* to 5 = *very often*). The measure consists of eight subscales that comprise three coping domains: approach, problem-focused avoidance, and emotion-focused avoidance coping strategies, which were developed based on research with healthy children and those with recurrent pain.³⁴ The measure has demonstrated good internal consistency with Cronbach's alphas for the subscales ranging from 0.78 to 0.89 and is considered "well-established".¹⁸ The Approach factor consists of information seeking, problem solving, seeking social support, and positive self-statement subscales and measures active coping methods of dealing with pain. The Problem-Focused Avoidance factor measures attempts to separate or disengage from pain and consists of positive self-statements, behavioral distraction, and cognitive distraction subscales. The Emotion-Focused Avoidance factor measures situations where emotions are freely expressed and there is limited effort to regulate feelings. In this sample, internal consistency ranged from 0.78 to 0.83.

Disease Severity.—At the end of the medical appointment, the SCD clinic nurse who conducted vitals and performed an initial health assessment on the child completed one 100mm VAS, with anchors 0 = *not at all* to 100 = *worst possible*, to assess the patient's disease severity. Previous research has demonstrated that health care provider ratings of disease severity via VAS are significantly correlated with objective ratings (i.e., hemoglobin levels and ER visits) in pediatric SCD.³⁵ In this sample, disease severity ratings provided by nurses were significantly positively associated with child report of pain, $r(101) = .26, p = .008$, which provides validity for using the item to estimate disease severity.

Demographic Information.—Accompanying caregivers completed a medical history and demographic form. Questions included child and caregiver age, child and caregiver races/ethnicities and genders, family income, and child's current disease and medication use.

Data Analysis Plan

Descriptive statistics, consisting of means, standard deviations, and percentages, were conducted to characterize the study sample and primary variables of interest (e.g., pain, pain-related coping, and HRQOL). Potential covariates (i.e., data collection site, child gender, child age, and disease severity) were identified through correlation and *t-test* analyses. Multiple mediation analysis was conducted to determine whether pain-related coping mediated the relation between child-reported pain and HRQOL. Multiple mediation can determine the extent to which a set of mediators (i.e., coping - Approach, Problem-Focused Avoidance, Emotion-Focused Avoidance), operating simultaneously, as well as individually, indirectly explains the relationship between the independent variable (pain) and dependent variable (HRQOL). Thus, we examined the (1) direct effects of pain on HRQOL (Hypothesis 1) and each coping type, (2) direct effect of each coping type on HRQOL, partialing out the effect of pain (Hypothesis 2), (3) total indirect effect of pain on HRQOL through all coping mediators together (Hypothesis 3), and (4) extent to which each coping mediator independently explained the relation between pain and HRQOL, accounting for the other variables in the model (Hypothesis 3). Multiple mediation analyses were conducted using the SPSS macro PROCESS model 4 (Hayes, 2013). The macro conducts bootstrapped unstandardized multiple linear regressions with 5,000 samples and provides estimates of the direct and indirect effects. Indirect effects are deemed significant when the 95% confidence intervals for the unstandardized path coefficients do not contain zero. Anticipating a small to modest effect size, Fritz and MacKinnon (2007) literature review and empirical analysis suggest that $N=115$ provides approximately 80% power ($\alpha=.05$, $\beta=.20$) for bootstrapped mediation models.³⁶ The final sample size of 104 participants approached recruitment goals and is similar to other studies examining HRQOL^{16,37,38} and pain-related coping models³⁹ in children with SCD. Covariates identified from preliminary analyses were included in the models. All statistical analyses were conducted using SPSS (22.0) for Windows (SPSS Inc., Chicago, IL, USA).

Results

Demographic Characteristics

Participants included 104 children 8 to 18 years of age ($M = 12.93$ years, $SD = 3.17$ years) with SCD. Recruitment occurred about equally across the two clinical sites (51.9% in one clinic). There were no significant child or caregiver demographic differences between the two sites of data collection. Fifty-five children were female (52.9%) and 96.2% were African American. One child was identified as Hispanic/Latino, one as Asian, and three caregivers did not identify child race. Caregivers reported that 72 (69.2%) children had hemoglobin SS, 13 (12.5%) had hemoglobin SC, 6 (5.8%) had Hemoglobin S-Beta Thalassemia, and 13 (12.5%) caregivers did not specify their child's subtype of SCD.

Most children were accompanied by a female caregiver (86.5%), which 84.6% were mothers. Caregivers ranged in age from 28 to 68 years ($M = 40.84$, $SD = 7.92$) and 96.2% self-identified as African American. Two caregivers were Hispanic/Latino, one was Native Hawaiian/Pacific Islander, and three did not report their race. Of the 95 caregivers who reported their annual family income, the median was \$40,001 to \$50,000. Fifty-nine

caregivers (56.7%) were married, 25 (24.0%) were single, and 19 (18.3%) were divorced or separated. Descriptive statistics of the main study variables are presented in Table 1.

Covariate Analysis

Preliminary covariate analyses with the main outcome variables revealed significant HRQOL differences between clinics, where children from the first clinic had significantly better scores on the Physical and Psychosocial subscales of the PedsQL (PedsQL Physical: $t(102) = 2.89, p = .005$, PedsQL Psychosocial: $t(102) = 2.14, p = .04$). This difference is likely due to more children ($n = 27$) from the second clinic compared to the first clinic ($n = 13$) attending a specialty clinic for increased SCD-related medical concerns, such as pulmonary complications. Site was included in the multiple mediation models as a covariate. No other differences were found between clinic sites on pain and pain-related coping variables.

T-tests revealed gender differences on the PedsQL Physical scale, $t(102) = 2.17, p = .032$, with males reporting significantly higher physical HRQOL compared to females. There were no other significant gender differences. Correlations between child age and disease severity and the primary variables of interest are presented in Table 2. Child age was significantly positively related to disease severity, $r(101) = .34, p = .001$, but not significantly associated with the main variables of interest (e.g., pain, pain-related coping, or HRQOL). Disease severity was significantly positively associated with child report of pain, $r(101) = .26, p = .008$ and significantly negatively associated with the PedsQL Physical scale (PedsQL Physical: $r(101) = -.23, p = .023$). Therefore, in addition to site, child gender and disease severity were included as covariates in the multiple mediation models.

Multiple Mediation Analysis

Physical HRQOL.—The multiple mediation model, including covariates (i.e., site, child gender, disease severity), accounted for 31.5% of the variance in Physical HRQOL.

Direct Effects (Hypotheses 1 & 2).—After controlling for covariates, the direct path of pain on Physical HRQOL was significant, $p < .001$ (Hypotheses 1; See Figure 1). The direct effect path from pain to Emotion-Focused Avoidance coping was approaching significance ($p = .086$) and the paths from pain to Approach and Problem-Focused Avoidance coping were not significant. The direct effect paths of Emotion-Focused Avoidance coping on Physical HRQOL, partialing out pain and controlling for covariates, was significant, $p = .018$ (Hypothesis 2). However, the direct effect paths of Approach and Problem-Focused Avoidance on Physical HRQOL, were not significant.

Total Indirect Effects (Hypothesis 3).—The total indirect effect of all the mediators operating in parallel on Physical HRQOL [pain $\rightarrow$ sum of mediators (Approach + Problem-Focused Avoidance + Emotion-Focused Avoidance coping) $\rightarrow$ Physical HRQOL] was not significant, Effect = $-.023$, Bootstrapped SE = $.022$, 95% CI = $-.078, .012$.

Specific Indirect Effects (Hypothesis 3).—The specific indirect effects of each coping mediator, while controlling for covariates and other coping variables in the model, were

examined to explore their unique indirect effects on Physical HRQOL. A significant mediation effect was found for Emotion-Focused Avoidance coping, Effect = $-.023$, Bootstrapped SE = $.018$, 95% CI = $-.0751$, $-.0003$, such that children reporting more pain were more likely to report engaging in more Emotion-Focused Avoidance coping strategies, which in turn was associated with reports of lower Physical HRQOL. Neither Approach nor Problem-Focused Avoidance coping mediated the relation between pain and Physical HRQOL.

Psychosocial HRQOL.—The multiple mediation model predicting Psychosocial HRQOL, including site, child gender, and disease severity as covariates, accounted for 39.5% of the variance in Psychosocial HRQOL.

Direct Effects (Hypotheses 1 & 2).—The direct path of pain on Psychosocial HRQOL was significant, $p = .032$ (Hypotheses 1), when controlling for covariates (See Figure 2). The direct effect paths from pain to the coping mediators were the same as above [i.e., Approach and Problem-Focused Avoidance not significant, Emotion-Focused Avoidance approaching significance ($p = .086$)]. The direct effect paths of Approach and Problem-Focused Avoidance coping on Psychosocial HRQOL, partialing out pain and controlling for covariates, were not significant. However, the direct effect path of Emotion-Focused Avoidance coping on Psychosocial HRQOL, partialing out pain and controlling for covariates, was significant, $p < .001$ (Hypothesis 2).

Total Indirect Effects (Hypothesis 3).—The total indirect effect of all the mediators operating in parallel [pain $\rightarrow$ sum of mediators (Approach + Problem-Focused Avoidance + Emotion-Focused Avoidance coping) $\rightarrow$ Psychosocial HRQOL] was not significant, Effect = $-.038$, Bootstrapped SE = $.025$, 95% CI = $-.0897$, $.0136$.

Specific Indirect Effects (Hypothesis 3).—No significant effects were found for Approach, Problem-Focused Avoidance, or Emotion-Focused Avoidance coping mediating the relation between pain and Psychosocial HRQOL.

Discussion

The purpose of this study was to examine the relationships between pain, pain-related coping, and HRQOL in children with SCD and specifically whether pain-related coping mediates the association between pain and HRQOL. Our hypothesis that pain would be associated with HRQOL was supported (Hypothesis 1). Specifically worst pain in the past week was associated with lower physical and psychosocial HRQOL. Our hypotheses that pain-related coping would be associated with HRQOL (Hypothesis 2) and would mediate the association between pain and HRQOL (Hypothesis 3) were partially supported. Specifically, we found emotion-focused avoidance coping, which consists of emotions being freely expressed with limited efforts to regulate feelings, was associated with decreased HRQOL and mediated the relationship between pain and physical HRQOL. However, these findings were not replicated with pain and psychosocial HRQOL or when approach coping and problem-focused avoidance coping were examined as mediators in the relationship between pain and HRQOL.

The finding that emotion-focused avoidance coping was related to increased pain and worse physical HRQOL in pediatric SCD is consistent with literature in other populations. Emotion-focused avoidance coping has been associated with increased pain intensity, distress, and functional disability in healthy pediatric populations and with symptoms of depression and anxiety in children experiencing recurrent pain.³² Our findings demonstrate the potential detrimental effects of emotion-focused avoidance coping on physical HRQOL and indicate that future interventions may benefit from integrating alternative coping strategies associated with reduced pain and HRQOL improvements. However, emotion-focused avoidance coping was not found to mediate the association between pain and psychosocial HRQOL. Thus, additional research examining pain, emotion-focused avoidance coping, and aspects of HRQOL in pediatric SCD is warranted.

We found that approach coping was negatively correlated with physical functioning, but did not serve as a mediator between pain and HRQOL. This is contrary to research demonstrating that approach coping is associated with improved overall functioning in youth¹⁹ and in other pediatric SCD samples.^{24,40} One possibility is that although current study participants reported practicing approach coping strategies, they may not be doing so frequently or effectively. Additionally, these types of coping strategies may not be as helpful for pediatric SCD patients not experiencing an acute pain crisis, such as those who participated in the current study. Problem-focused avoidance coping was also not found to be associated with pain or dimensions of HRQOL in this sample. This finding is somewhat inconsistent with previous literature, which has found that problem-focused avoidance coping is associated with decreased pain and distress in pediatric populations.^{19,34} The problem-focused avoidance scale from the PCQ was significantly correlated with the approach coping scale but not with the pain or HRQOL variables. It is possible the lack of statistical findings may be due to children in this study reporting an overall lack of engagement in the coping strategies making up this specific scale (i.e., positive self-statements, behavioral distraction, and cognitive distraction subscales). To confirm our findings regarding problem-focused avoidance coping in pediatric SCD, future research may benefit from utilizing different assessments of pain-related coping that examine additional dimensions of coping, as well as more comprehensive assessments of disease-related pain, such as daily diaries.

Generally, our findings are consistent with the risk and resistance model of adjustment.^{23,41} Specifically, the results from this study suggest that emotion-focused avoidance coping may be considered a risk factor in the model due to negative impacts on pain and physical HRQOL. Continuing to examine the utility of the risk and resistance model, as well as other potential models, to conceptualize adjustment in pediatric SCD is vitally important to inform future research and clinical practice.

This study had several important strengths. First, this study examined multiple dimensions of pain-related coping and examined them as mediators in the association between pain and HRQOL, which provided unique information regarding pain-related coping strategies that may be beneficial for children with SCD. The findings from this study also point to the importance of continuing to identify pain-related coping strategies that are beneficial in pediatric SCD to inform the development of future interventions focused on pain

management and improved HRQOL. A second strength of this study is our moderate sample size, particularly when compared to previous studies focusing on pediatric SCD with primarily small sample sizes. Third, the use of multiple mediation and bootstrapping analyses was a strength of the current study as it allowed us to provide a more robust statistical examination regarding associations between pain, pain-related coping, and HRQOL compared to prior research and provides support for longitudinal studies to examine the temporal relations between these constructs in children with SCD. Finally, recruiting participants from multiple clinical sites in the same urban area makes our findings more generalizable to children receiving care for SCD in this part of the country.

Despite the strengths identified, this study also had limitations. Though we recruited participants from multiple clinics, future research needs to be conducted with larger samples and in other geographical locations to determine if our results are regionally or internationally generalizable. Additionally, the majority of families in this sample were middle income and married, which may not be representative of children with SCD across the U.S.^{42,43} Children participating in this study were attending a regularly scheduled SCD-related medical visit. Children with SCD in this study reported somewhat lower worst pain in the past week than expected, which limited the range of pain values and potentially our ability to detect associations. Therefore, findings from this study may not generalize to children with SCD experiencing acute pain crises or attending non-routine medical visits. In future research current findings would be augmented by the examination of pain, pain-related coping, and daily functioning (e.g., HRQOL, functional disability) using additional measures of pain, including measures of disease severity, history of vaso-occlusive pain episodes, and pain-related coping, as well as while children are receiving treatment for an acute pain crisis. Further investigation of how specific pharmacologic (e.g., hydroxyurea) and psychological (e.g., cognitive-behavioral therapy) pain management interventions relate to HRQOL and pain-related coping may also be helpful. Although we controlled for age in analyses, another potential limitation is the broad age range of our sample. Given the increased frequency and intensity of pain from childhood to adolescence with SCD,⁴⁴ it is possible that adolescents may have different disease-related pain experiences and utilize different coping styles than their younger counterparts. Thus, future research would benefit from examining associations between pain, pain-related coping, and HRQOL in children and adolescents separately to determine whether developmental considerations should be incorporated into psychological interventions. Finally, this study was cross-sectional in nature. Following children with SCD over time utilizing daily diaries and repeated time points would strengthen our evidence regarding the mediating role of pain-related coping in the association between pain and HRQOL.

Additional research is needed to understand the mechanisms through which pain-related coping influences the association between pain and HRQOL in pediatric SCD. For example, interpersonal factors, such as feelings of competence related to managing pain and self-esteem, which have been identified in the risk and resistance model of adjustment²³ may be important mechanisms that could influence coping strategies utilized and also impact emotional and behavioral functioning that may influence HRQOL. Examining pain-related coping and additional functioning outcomes, like functional disability, in children with SCD would also be warranted. Given that SCD primarily impacts African Americans, it is

especially important to consider culturally sensitive measures of pain-related coping, as well as culturally tailored psychological interventions to help SCD children and adolescents cope with pain. For example, spiritual and religious related coping strategies may be an important factor that influences HRQOL in this population.⁴⁵ This line of research could be used to evaluate the applicability of the biopsychosocial-spiritual model of chronic pain that has been proposed in adults with SCD⁴⁶ and identify culturally appropriate and acceptable pain coping strategies that could be incorporated into future interventions to improve pain and HRQOL in pediatric SCD.

In summary, we found partial support for our hypotheses. Emotion-focused avoidance coping was related to worse pain and in turn decreased physical HRQOL. Emotion-focused avoidance was the only type of pain coping that mediated the relationship between pain and physical HRQOL in this sample of pediatric patients with SCD. Future research should continue to explore associations between pain, pain-related coping, and HRQOL longitudinally in this population, as well as examine ways to reduce the use of emotion-focused avoidance coping if our findings are confirmed. Teaching patients alternative methods of pain-related coping that may improve pain and HRQOL related outcomes would be of importance.

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Figure 1.

Numbers represent path coefficients of the direct effects. Clinic Site, Child Gender, and Disease Severity (i.e., nurse-report) were controlled for as covariates in this model. Dashed lines indicate non-significant associations. † $p < .10$, * $p < .05$, *** $p < .001$. 95% Confidence intervals (CI) not containing zero indicate significant mediating indirect effects, which if present, are indicated in Bold above.

Figure 2.

Numbers represent path coefficients of the direct effects. Clinic Site, Child Gender, and Disease Severity (i.e., nurse-report) were controlled for as covariates in this model. Dashed lines indicate non-significant findings. [†] $p < .10$, * $p < .05$, *** $p < .001$. 95% Confidence intervals (CI) not containing zero indicate significant mediating indirect effects, which if present, are indicated in Bold above.

Table 1

Descriptive Statistics of Main Study Variables (N = 104)

Measures	M	SD	Range
Pediatric Pain Questionnaire (PPQ) Worst Pain ^a	29.46	34.57	0 – 100
Pain Coping Questionnaire (PCQ) ^b			
Approach	3.05	0.61	1.58 – 4.37
Emotion-Focused Avoidance	1.87	0.62	1.00 – 3.50
Problem-Focused Avoidance	2.80	0.79	1.00 – 4.90
Pediatric Quality of Life Inventory (PedsQL) ^c			
Physical Score	69.46	20.96	15.63 – 100
Psychosocial Score	71.07	16.46	28.57 – 100

Note.

^aScores on the PPQ range from 0 to 100, with higher scores representing more pain.^bScores on the PCQ range from 1 to 5, with higher scores representing more frequent use of the coping strategy.^cScores on the PedsQL range from 0 to 100, with higher scores representing higher quality of life.

Table 2

Correlations between Child Age and Disease Severity and Main Study Variables

	1.	2.	3.	4.	5.	6.	7.	8.
1. Child Age	-							
2. Disease Severity	.34***	-						
3. PPQ Worst Pain	.12	.26**	-					
4. PCQ Approach	.01	.10	.15	-				
5. PCQ Problem-Focused Avoidance	-.17	.01	.09	.24*	-			
6. PCQ Emotion-Focused Avoidance	-.01	.07	.18 [†]	.16	-.02	-		
7. PedsQL Physical	-.06	-.23*	-.43***	-.20*	.04	-.32***	-	
8. PedsQL Psychosocial	.08	-.17	-.32**	-.11	.01	-.58***	.69***	-

Notes.

PPQ = Pediatric Pain Questionnaire; PCQ = Pain Coping Questionnaire; PedsQL = Pediatric Quality of Life Inventory.

[†]
 $p < .10$ *
 $p < .05$ **
 $p < .01$ ***
 $p < .001$.