A Family-Based Cognitive-Behavioral Intervention for Pediatric Patients with Sickle Cell Disease

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Abstract

Background: The purpose of this study was to examine the impact of a culturally sensitive, cognitive-behavioral family treatment (CBFT) for pediatric patients with Sickle Cell Disease (SCD) to improve pain symptoms, health-related quality of life, functionality, depression, and coping strategies. Individual cognitive-behavioral treatment has been shown previously to be effective at improving pain symptoms, functionality, adaptive coping, and health care utilization, but such benefits have not yet been shown for SCD patients. The present study aimed to address this limitation by modifying the intervention to both include the family and to utilize culturally sensitive practices, which may be particularly relevant for this population. **Methods:** A non-concurrent multiple baseline design was used to assess the effectiveness of the intervention. A sample of 4 children (ages 8 to 12) and 4 adolescents (ages 13 to 15) participated in the intervention. Manualized treatment consisted of five sessions (including child and parent) that targeted problem-solving skills, cognitive processes, coping strategies, goal setting, and family processes. Outcomes of interest including health-related quality of life, functionality, psychological adjustment, and coping strategies, were assessed by child and parent report at pre-treatment (baseline), post-treatment, and 2-, 4-, and 6-month follow-up. Participants completed daily diaries to quantify pain, anxiety, and functionality. **Results:** Repeated-measures general linear model analyses were run separately for all outcome variables. A significant main effect of time was found for youth-reported HRQoL, $F(4, 20) = 4.6$, $p=.01$, depressive symptomatology, $F(4, 20) = 4.5$, $p=.01$, and parent-reported Internalizing, $F(4, 16) = 3.4$, $p=.03$, Externalizing, $F(4, 16) = 7.2$, $p=.00$, and Total Behavior Problems, $F(4, 16) = 7.7$, $p=.00$ from baseline to 6-

month post-treatment. The mean frequency of pain symptoms also decreased for five of the eight participants (i.e., visual inspection of the daily diaries from baseline to treatment). **Conclusions:** These results suggest the potential for clinical gains through the incorporation of culturally sensitive and family-based practices into existing cognitive-behavioral interventions for SCD. The symptomatic improvements observed in the present study indicate gains in both specific domains (i.e., pain), as well as general psychological outcomes (i.e., improvements in depression, health-related quality of life, internalizing and externalizing behaviors).

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"Focus on the journey, not the destination. Joy is found not in finishing an activity but in doing it." (Greg Anderson, 1964)

While it is hard to believe that this journey is nearly complete, the challenging times and lessons learned throughout were well worth it. Although, in moments doubts were surely felt, it was during those times that I learned about myself and the people around me.

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Introduction

Over the past decade there has been an increased interest in the psychosocial adjustment and quality of life of children with chronic illness. In the pediatric chronic illness literature, Sickle Cell Disease (SCD) has received less attention among interventions targeting pain secondary to chronic illness. As a chronic health and recurrent pain condition, SCD impacts many domains of functioning and thus requires considerable adjustment to the limitations, stressors and associated symptoms of the disease. Quality of life and functional status are most often utilized to characterize adjustment to SCD in pediatric populations (Palermo, 2000). Research findings indicate that children with SCD experience variable profiles of adjustment, with some subsets exhibiting normal adjustment profiles, while others miss considerable school days, have limited social and athletic activities, and experience emotional distress and poor quality of life (Palermo, Witherspoon, Valenzuela, & Drotar, 2004). In addition to health-related stressors, children and adolescents with SCD are faced with challenges related to ethnic minority status and the associated experiences of prejudice, discrimination, decreased access to health care, urban environmental stressors, and barriers related to socioeconomic status (SES) that may have an impact on their health status and psychosocial adaptation (Barakat, Lash, Lutz, & Nicolaou, 2006).

Many children with SCD also experience depression, anxiety, or peer and interpersonal problems, which may be due in part to their pain (Gil, Abrams, Phillips, & Keefe, 1989; Thompson, Gil, Burbach, Keith, & Kinney, 1993a). Children's coping responses to pain (i.e., the behaviors that children use to deal with pain) have been identified as a likely contributor to the

wide variability in outcomes in children with chronic and recurrent pain. A child's coping strategies for pain, stress appraisal, and mother's psychological adjustment predict (parent- and child-report of) child psychological adjustment outcomes (Thompson et al., 1993a,b). Longitudinal assessment studies have further indicated that child coping strategies are less stable over time and thus possibly more amenable to change (Gil et al., 1997), suggesting the benefit of early interventions that target these coping strategies.

In fact, coping strategies account for a significant portion of variance in pain report and psychosocial and functional adjustment in patients with SCD, even after controlling for demographic and disease severity (Gil, Anthony, Carson, Redding-Lallinger, Daeschner, & Ware, 2001). Research is currently focused on targeting and testing psychosocial interventions, focusing specifically on cognitive-behavioral techniques (which have been shown to be more beneficial than behavioral and social support intervention), to improve SCD pain, quality of life, and health outcomes (Chen, Cole, & Kato, 2004). Consistent with this research aim, the objectives of Healthy People 2020 (Department of Health and Human Services [DHHS], 2011) include reducing hospitalizations due to preventable complications of SCD, improving quality of life and increasing activity involvement.

Characteristics of Sickle Cell Disease (SCD)

SCD is a group of inherited disorders affecting approximately 1 in every 400-500 African American newborns in the United States (Tarnowski & Brown, 2000), making it the most prevalent genetic conditions in the United States (Agency for Health Care Policy and Research, 1993). The disorder primarily affects people of Caribbean and African origin, but also affects a small percentage of people of Indian, Mediterranean and Middle Eastern descent (Swain,

Mitchell, & Powers, 2006). SCD is also prevalent in Hispanic and Latino populations. SCD results from an autosomal recessive genetic deficit and is classified by genotype. Individuals affected with SCD demonstrate abnormal genes for hemoglobin (Hb) S, which produces a change in the shape of red blood cells from their normal disk shape to a sickle shape. These abnormally shaped cells obstruct normal blood flow and production of new red blood cells. Of the three sickle cell syndromes, the most common is the homozygous condition, *sickle cell anemia* (Hb SS), which is caused by two abnormal genes for hemoglobin S and is associated with earlier, more frequent and severe symptoms (Charache, Lubin, & Reid, 1989). The sickle cell anemia condition occurs in about 1 in 500 African American births. *Sickle cell hemoglobin C* (Hb SC) is caused by the hemoglobin target cells combining with the sickle hemoglobin S; *Sickle β-thalassemia* (Hb S β-thalassemia) involves the inheritance of both the thalassemia and sickle cell genes. Both sickle cell hemoglobin and sickle β-thalassemia produce a milder degree of symptoms and complications. The sickle cell hemoglobin condition occurs in about 1 in 835 African American births, while the Hb S β-thalassemia occurs in approximately 1 in 50,000 births (Ievers-Landis, Brown, Drotar, Bunke, Lambert, & Walker, 2001). The distinction between the sickle cell syndromes is based on the source of genetic abnormality, rather than an identifiable difference in the symptom profile of each. Any of the types may be characterized by severe pain symptoms and variability in symptom profile.

Symptoms of SCD include chronic anemia, susceptibility to infection, and vaso-occlusive crises or vaso-occlusive episodes (VOEs), which result in severe pain that can last hours to weeks (Beyer, Simmons, Woods, & Woods, 1999; Murray & May, 1988). This vaso-occlusion is caused when sickle cells are unable to flow through arteries, capillaries, arterioles, and other blood vessels and as a result, obstruct blood flow. This sickling can occur anywhere in the body,

including fingers, arms, ribs, abdomen, and organs such as the brain and eyes (Swain et al., 2006), but it most commonly occurs in the spleen, bones, and joints (Elander & Midence, 1996). Vasoocclusions that occur in the brain put children with SCD at increased risk for neurological disease, including stroke. Cerebrovascular accidents occur in 5-10% of children with SCD, and "silent" strokes occur in 11-20% of children with SCD (Balkarab et al., 1992).

The course of SCD during the lifespan is highly variable; it can change within one person over time and/or be quite different from that of another person. The primary characteristic of SCD is the unpredictable, acute painful crises. VOEs may or may not be preceded (or triggered) by certain events such as illness, dehydration, trauma, exposure to cold, emotional upset (Swain et al., 2006). Pain from a VOE can be the most severe symptom and is the most common cause of acute morbidity in SCD (Shapiro, 1993). However, one third of patients with SCD do not experience VOEs serious enough to warrant medical treatment. Their episodes may be frequent, however, resulting in significant absences from school and peer group activities which place them at academic and psychosocial risk (Walker & Jacobs, 1984). Two thirds of patients with SCD will experience VOEs multiple times per year, with associated moderate to severe pain (Shapiro, 1993).

While the majority of SCD hospital admissions (approximately 90% of all SCD-related emergency admissions) (Swain et al., 2006) are because of VOEs, the majority of painful episodes, nearly 90% in one study of children and adolescents with SCD, are treated and managed in the home environment (Shapiro et al., 1995). Most often, only severe pain episodes require hospitalization; therefore, mild-to-moderate painful episodes and chronic SCD pain are subject to home management (Ballas, 2002). In other words, families appear to bear the primary burden for managing the child's pain episodes. In contrast, mild pain is usually treated with

nonpharamacological agents alone (i.e., bed rest, hydration, massage, relaxation, heating pads, baths) (Jacob, Miaskowski, Savedra, Beyer, Treadwell, & Styles, 2003) or in combination with a nonopioid medication.

Current treatment focuses primarily on the management of the chronic and repetitive VOEs. As such, treatment for SCD varies in intensity and invasiveness depending on severity of complications. It may involve daily management (i.e., hydration, restrictions on activities, prophylactic antibiotics, and pain management) as well as preventative follow-up care. In addition, for children who have had stroke, are at risk for stroke, or experience severe pain crises, regular blood transfusions are required. For those children who experience the most severe symptoms of SCD (i.e., characterized by frequent, intense pain crises or VOEs) hydroxyurea, the only FDA approved drug for treatment of SCD, appears to be an effective prophylactic agent that reduces the frequency of painful episodes (VOEs), acute chest syndrome, transfusion requirement, and decreases mortality (Amrolia, Almeida, Davies, & Roberts, 2003; Raghupathy & Billet, 2009). However, one third of patients treated with hydroxyurea remain unresponsive to the drug (Raghupathy & Billet, 2009). For these most severely affected patients there are limited options for additional medical treatment, placing them at risk for adverse adaptive outcomes, impacting their quality of life and functional status.

Indices of Child Psychosocial Adjustment

The concept of psychosocial "adjustment" is multidimensional and should not be equated with the uni-dimensional notion of reduced or eliminated pain as the measure of success. As a result, successful adjustment can be measured across various affective, behavioral, and perceptual domains. Adjustment factors most relevant for pediatric patients are pain severity,

pain behaviors, activity level, health care utilization, anxiety and depression (Jensen, Turner, Romano, & Karoly, 1991). Although these adjustment parameters have been characterized in many different ways in the literature, in the present study adjustment outcomes will be quantified along the following domains: *pain*, *health-related quality of life*, *functionality*, and *psychological adjustment*. It should be noted that there is considerable overlap inherent in these dimensions – any change in one domain impacts outcomes in other domains (Bronfenbrenner, 1977); however, the distinction made here is intended to capture more specific features of a child's broader adjustment. *Pain* outcomes characterize the frequency and duration of physical symptoms. *Health-related quality of life* encompasses general life satisfaction and happiness. *Functionality* describes limitations in typical daily life activities, which for pediatric patients may be most easily characterized by examining the number of school absences and health care contacts. *Psychological adjustment* captures outcomes including behavior problems and other psychopathology.

Chronic and Recurrent Pain. "Pain" can be defined as an unpleasant experience that accompanies both sensory and emotional modalities, and is influenced by multiple factors, including cognitive, affective, and environmental (Merskey & Bogduk, 1994). For recurrent pain in particular, which is episodic (usually brief) and extends over a long period of time, social and behavioral factors may be have more influence on illness behavior (Turk & Okifuji, 2001). Chronic and recurrent pain frequently impact everyday functioning of children in significant ways. For example, children whose chronic pain limits their functioning have an increased risk for lifelong problems with pain, as well as psychological disturbance (e.g., anxiety, depression) in adulthood (Palermo, 2000). Sickle cell pain is complex and is strongly affected by multiple factors including pathophysiological, psychological, social, cultural, and spiritual factors (Ballas,

2002). Recurrent pain, anemia, low exercise tolerance, lung problems, growth delays, strokes, priapism, delayed sexual maturation, and enuresis can accompany SCD to further complicate adjustment outcomes (Hoppe, Styles, & Vichinsky, 1998; Steinberg, 1999). Of all possible complications, the frequency of pain events in children has been found to result in the most significant morbidity and mortality in adulthood (Platt et al., 1991).

Because pain is central and a common complication of SCD, learning to manage pain effectively is essential to maintaining an active lifestyle and optimal quality of life. Developing effective pain management strategies by adolescence seems to allow patients with SCD to manage the transition to adult services more effectively and to become more competent adult patients (Benjamin et al., 1999). Pain and other complications can have a significant impact on quality of life for African American youths living with SCD when compared with African American youths living without SCD (Palermo, Schwartz, Drotar, & McGowan, 2002).

Health-Related Quality of Life. The advent of more aggressive methods of treating pediatric chronic illness and the success of these treatments in prolonging life expectancy has led to concerns about the impact of treatment on psychosocial and functional outcomes or quality of life (QoL) (Barakat, Lutz, Smith-Whitley & Oheme-Frempong, 2005). Health-related quality of life (HRQoL) refers to the specific impact of an illness or medical treatment on an individual's physical, cognitive, social, and emotional functioning (Grootenhuis, Koopman, Verrips, Vogels, & Last, 2007). For example, the effects of SCD and its treatment often increase the child's dependence on his or her parents and decrease their participation in peer- and school-based activities (Grootenhus, Koopman, Verrips, Vogels, & Last, 2007). Furthermore, the lack of predictability of pain episodes may result in decreased adherence to treatment when children feel physically well (La Greca & Bearman, 2001). For children with more severe and persistent

complications, the demands of rest and hydration, routine follow-up medical visits, and hydroxyurea may seem daunting and yet acceptable; these children may view treatment as necessary to improve physical and psychosocial functioning (Barakat et al., 2005). Alternatively, for children with SCD who have few complications the demands of treatment may seem inordinate, and they may therefore engage in fewer SCD-related care activities in order to maintain or improve their HRQoL (Barakat et al., 2005). HRQoL is an important construct to measure in children with SCD to describe their health profile and functional status, to evaluate the effects of new medical treatments, and to evaluate their needs and those of their families. Despite this need, there are limited data and inconsistent findings on HRQoL in children with SCD.

Functional Impairment. For children with chronic and recurrent pain, their daily lives may be impacted by many functional limitations (lowered levels of happiness and satisfaction with life, fewer healthy days) that often result from pain episodes, which in turn result in higher emotional distress and poor quality of life (Palermo et al., 2004). Functional impairment captures difficulty in performing age-appropriate physical, mental, and social activities in daily life due to physical health status. The functional impact of chronic and recurrent pain on children and adolescents can be substantial. School-related functional impairments have been most often documented in studies of pediatric SCD, including frequent school absences and limitations in social and athletic activities (Fuggle, Shand, Gill, & Davies, 1996; Palermo, 2000; Palermo et al., 2004). For example, Shapiro and colleagues (1995) found that children with SCD who experienced frequent pain were absent from school on 21% of school days (6-8 weeks per year). Adolescents evaluate their quality of life as less satisfactory than that of their healthy peers,

primarily as a result of the daily frustrations of chronic pain, including frequent hospitalization, school absences, social isolation, daily stress, and lowered self-esteem (Merlijn et al., 2005).

Psychological Adjustment. In comparison with demographically matched comparison groups, children and adolescents with SCD are at risk for the development of psychological distress and problems in the areas of internalizing disorders and self–esteem. Children and adolescents with SCD appear to be at most risk for internalizing problems (particularly anxiety-based disorders), and may not be at any increased risk for externalizing behavior problems (Barakat et al., 2006). Investigations have found estimates of psychological problems occurring in one third to almost two thirds of samples (Barbarin, Whitten, & Bond, 1994; Thompson et al., 1993a; Thompson, Gustafson, & Gil, 1995), although the findings with respect to the specific risk for psychopathology are quite mixed. On the one hand, research has shown that children with SCD have increased behavior problems and psychopathology (Kell, Kliewer, Erickson, & Ohene-Frempong, 1998; Thompson et al., 1993a; Thompson, Gil, Keith, Gustafson, George, & Kinney, 1994), lowered self-esteem (Charache et al., 1989; Hurtig & White, 1986), less adequate interpersonal interactions and social competence (Hurtig & White, 1986; Noll, Vannatta, Koonz, Kalinyak, Bukowski, & Davies, 1986), and problematic family functioning (Nevergold, 1987). In contrast however, other studies report that the majority of children and families successfully adapt and cope with the challenges associated with SCD (Barbarin & Christian, 1999; Lemanek, Moore, Gresham, Williamson, & Kelley, 1986; Midence, McManus, Fuggle, & Davies, 1996). These discrepant outcomes may in part be attributed to widely diverging methodologies, however it is likely that it is also a function of the inherent variability of SCD symptomatology.

However, good adjustment for children and adolescents with SCD is also possible. Findings from the Cooperative Sickle Cell Disease study (Thompson, Armstrong, Link,

Pegelow, Moser, & Wang, 2003) point to the remarkable resilience of many children and adolescents with SCD in that problems noted are often not at a level of psychopathology, and that youths with SCD may function adaptively. This provides a reminder that strength can be developed while facing difficult stressors, and that children and families can thrive in the face of such challenges. In addition, these findings suggest the likelihood of intervening in less well-adapting families in order to increase the chances of thriving under difficult circumstances. Overall then, pain, HRQoL, functionality and psychological adjustment serve to capture the extent to which chronic pain has a daily impact on the lives of pediatric patients, and as such, will be used as a means of quantifying treatment gains in the present study.

Conceptual Framework for Adaptation to Sickle Cell Disease

From an ecological perspective, understanding adaptation to childhood chronic illness requires examining contextual factors that extend beyond the expected physical manifestations to complex, reciprocal influences among the illness and its treatment, the child, and the family. As such, research on child adaptation to chronic illness has shifted from focusing on isolated factors associated with adjustment to a specific illness to more conceptually based and model driven research to identify the multitude of factors associated with psychosocial adjustment and how they interact (Casey, Brown, & Bakeman, 2000).

There are two major models that have been proposed: the risk-resistance model (Wallander & Varni, 1992), and the transactional stress and coping (TSC) model (Thompson & Gustafson, 1996). The utility of these two theoretical models has been demonstrated in a large number of studies on the psychosocial adjustment of children, adolescents, and adults (Burlew, 2002). Both theoretical models imply that psychosocial factors might serve as potential

protective mechanisms by buffering the impact of the stressor (e.g., medical severity) on adaptation. The existing body of research associated with these models has made a substantial contribution by identifying psychosocial variables associated with variability in adaptation. In the risk-resistance model, both risk and resistance variables mediate and moderate the relation between disability stress and adjustment. The complex interplay between these risk factors, such as illness parameters (e.g., diagnosis and severity), functional independence (e.g., functional limitations from the disease) and psychosocial stressors, and resistance factors (such as stress processing, intrapersonal characteristics and social ecological factors) influence a child's adjustment to a chronic illness (Hocking & Lochman, 2005).

In the present study, the TSC model will provide the theoretical framework for exploring child and family adaptational processes related to adaptive outcomes for pediatric patients with SCD. This choice is based upon several considerations. First, the TSC model was developed from the risk-resistance model in order to more specifically identify child and family parameters of adaptation. As such, it more closely maps onto the processes of interest in the present study by directly targeting the individual cognitive processes, coping strategies and family characteristics believed to affect outcomes in the SCD population. Some of the variables proposed in the risk-resilience model (i.e., functional independence) are characterized as predictors of outcome, whereas more recent research has identified that they may be more appropriately considered as outcome variables. Finally, the TSC model serves as a more parsimonious framework for capturing adaptational child and family processes in understanding adaptive outcomes for the SCD population.

The TSC model (see figure 1) outlines the multilevel process of adjustment to chronic illness (Thompson et al., 1993; Thompson, Gustafon, Hamlett, & Spock, 1992; Thompson et al.,

1994). From this model, SCD is a potential stressor to which the pediatric patient and family must adapt. Adjustment to the illness is not simply the direct function of the illness, but is mediated by transactions or exchanges between illness parameters (illness type and severity), demographic parameters (child's age, gender, and SES), and child and parent adaptational processes (appraisal, self-efficacy, coping). The TSC model is guided by the work of Lazarus and Folkman (1984) which describes three types of psychosocial influences on child adjustment to chronic illness: (1) *cognitive processes*, including stress appraisal (Lazarus & Folkman, 1984), self-efficacy (Bandura, 1988) and health locus of control (Strickland, 1978); (2) *coping strategies* (Lazarus & Folkman, 1984); and (3) quality of *family processes* (Moos & Moos, 1981). Research has demonstrated that these processes serve to reduce the impact of stress and have saliency as targets for potential interventions (Hocking & Lochman, 2005; Thompson et al., 1993b). Variables consistent with this model have been demonstrated to account for 30% to 68% of the variance in psychosocial adaptation among children with SCD (Burlew, Telfair, Colangelo, & Wright, 2000). For these reasons, the general framework of the TSC model has guided the selection of parent and child adaptational processes that will be targeted by the pain intervention in the present study. The primary components of the TSC model are discussed below, including demographic characteristics, cognitive processes, coping strategies and family processes.

Demographic Characteristics. Developmental differences have been reported to play a role in adaptation to SCD. Adolescents with SCD generally experience more problems in adaptation than their younger counterparts (Brown, Eckman, Baldwin, Buchanan, & Dingle, 1995; Hurtig, Koepke, & Park, 1989), although these findings have not been consistently reported (Lutz, Barakat, Smith-Whitley, & Ohene-Frempong, 2004; Thompson et al., 2003).

Adolescence is a critical time developmentally in terms of separation from the family, the formation of one's identity, and the centrality of social relationships. SCD, in combination with risk factors associated with lower SES and minority racial status, present an additional set of challenges for adolescents with SCD, which may impede their ability to gain independence from the family and to find acceptance from peers, both of which are essential to transverse adolescence (Baskin et al., 1998). Adolescents with SCD report more problems with peer relationships, greater behavior problems, and poorer self-esteem compared with health peers and with younger children with SCD (Scott & Scott, 1999). Moreover, nonadherence to treatment recommendations increases during adolescence partly because of more adaptation problems, increased responsibilities for medical care, and developmental demands particular to adolescence (Barakat, Smith-Whitley, & Ohene-Frempong, 2002; Baskin et al., 1998).

Gender differences present a second qualifier to adaptational outcomes. Males with SCD, particularly in adolescence, experience greater problems than females in adaptation, including internalizing symptoms, social relations, and low self-esteem (Brown et al., 1995; Brown, Kaslow, et al., 1993; see Baskin et al., 1998 for a review; and see Thompson et al., 2003 for an exception). These findings may be attributed to higher SCD complications in males (Barakat et al., 2002; Lutz et al., 2004) and the impact that SCD complications have on separation and individuation from the family, identity development, and social acceptance and support (Scott & Scott, 1999); males with SCD reported poorer family functioning than females, thus supporting this interpretation (Lutz et al., 2004). For example, short stature, delayed puberty, and decreased physical stamina are more likely to have a negative impact on male adolescents' successful transition through adolescence than that of females because of the more central role that physical prowess plays in adaptation of male adolescents (Barakat et al., 2006). In addition, the cultural

emphasis on male strength and physical activity place adolescent males with SCD at an increased risk for maladaptive outcomes (Hurtig & White, 1989).

Cognitive Processes. The relevance of cognitive processes of stress appraisal, expectations of efficacy, and locus of control has been well established with regards to adjustment (Bandura, 1988; Lazarus & Folkman, 1984). With regards to SCD there are two pivotal sources of stress. One source is associated with chronic illness, including dealing with the symptoms and treatments, maintaining emotional well-being, and preparing for an uncertain future (Thompson et al., 1993b). Another source of stress comes in the form of daily hassles that stem from everyday transactions with the environment (Thompson et al., 1993b).

Appraisal of stress captures how the patient conceptualizes the subjective evaluation of stressors, that is, the individual's evaluation of which coping options are available, the likelihood that a given coping option will accomplish what it is supposed to, and the likelihood that one can apply a particular strategy effectively. Stress appraisal has been associated with adjustment in children with SCD (Gil et al., 2003; Thompson, Gustafson, Gil, Godfrey, & Bennet-Murphy, 1998). Using a daily diary method, Gil et al. (2003) found that increases in perceived stress were associated with same day increases in pain, health care use, and reduced school and social activity in adolescents with SCD. Thompson and colleagues (1998) also found that children's appraisal of stress accounted for a significant amount of the variance in adjustment outcomes. Lewis and Kliewer (1996) examined the effect of hope on physical and psychological adaptation of children and adolescents with SCD. The results indicated that higher levels of hope were associated with lower anxiety symptoms, lower physical anxiety symptoms, and higher concentration.

Self-efficacy beliefs, or beliefs about the ability to manage or function with pain, affect the initiation, strength, and persistence of coping behavior (Arnstein, Caudill, Mandel, Norris, & Beasley, 1999; Bandura, 1988; Coughlan, Ridout, Williams, & Richardson, 1995; Thompson et al., 1993b). Arnstein et al. (1999) found that self-efficacy (related to pain management, coping, and physical functioning) mediated pain intensity and disability, and that both self-efficacy beliefs and pain intensity contributed to depression. Coughlan and colleagues (1995) reported higher drop-out from a pain management program when self-efficacy beliefs were not improved by treatment. Internal locus of control expectations have also been associated with less psychological distress and more adaptive responses to health problems (Thompson et al., 1993b). Adjustment was examined in a study of 55 children (age 5 to 16 years) with SCD and their primary caregivers (Brown, Lambert, et al., 2000). In terms of parent report of child adjustment, 20% met criteria for poor adjustment in terms of internalizing problems (e.g., symptoms of anxiety, depression), and 35% met criteria for poor adjustment in terms of externalizing problems (e.g., acting-out behaviors). Only child health locus of control emerged as a significant predictor of child internalizing and externalizing problems. Children's reports of a cognitive processing style characterized by internal beliefs of expectation of control over their health were associated with better child adjustment (Brown et al., 2000).

Coping. Coping strategies (i.e., the ways in which people deal with stress) have also been linked to adjustment. Coping efforts encompass all attempts to deal with the multiple physical, emotional, and behavioral ramifications of a chronic pain condition. Importantly, for patients with chronic pain, the pain stimulus itself is not the only stressor with which they must cope, and various stressors may require potentially different coping efforts. The use of passive coping strategies (regulating emotional states that are associated with, or result from stress, generally

involving orientation away from the stressor, e.g., self-isolation, catastrophizing, and disengagement) is associated with poorer adjustment to chronic illness including higher levels of pain, depressive symptoms, and functional disability in children and adolescents with SCD (Reid, Gilbert, & McGrath, 1998; Thompson et al., 1993b; Walker, Smith, Garber, & Van Slyke, 1997; Walker, Smith, Garber, & Claar, 2005). In contrast, the use of active coping strategies (efforts to accept or adapt to the stressor, e.g., acceptance, minimizing pain, self encouragement, and distraction) are associated with both lower initial pain reports, decreases in pain over time, and lower levels of anxiety and depressive symptoms (Lazarus & Folkman, 1984; Thompson et al., 1993b; Walker et al., 1997). In particular, research by Lewis and Kleiwer (1996) indicated that greater use of active coping was related to lower levels of physiological anxiety symptoms, while increased distraction coping was associated with more symptoms of depression. Finally, more reliance on avoidance coping strategies was associated with difficulties with concentration and anxiety symptoms.

For children with SCD, strategies for coping with pain characterized by negative thinking and passive adherence have been associated with more psychological distress, functional impairment, and health care use (Gil, Williams, Thompson, & Kinney, 1991). In a sample of 35 adolescents (13-17 years old) with SCD, adolescents with poor adjustment not only had significantly lower expectations of efficacy regarding illness tasks, but also higher levels of palliative coping, and pain-coping strategies characterized by negative thinking and passive adherence (Thompson et al., 1995). After controlling for other variables in the TSC model, children's pain-coping strategies accounted for a significant amount of the variance in child-reported symptoms (19%) (Thompson et al., 1994, 1995). These findings suggest that decreasing

pain-coping strategies characterized by negative thinking may be helpful in efforts to enhance adjustment in children with SCD (Thompson et al., 1994).

Spirito and colleagues (1995) examined coping strategies in 177 children and adolescents with chronic illness, including SCD. Results indicated that adolescents used strategies such as blaming others and wishful thinking less than children; however, adolescents engaged in higher levels of resignation. For children and adolescents with SCD, coping strategies have been related to the extent of health care utilization, in that, active coping has been associated with reduced health utilization, including emergency room visits (Gil et al., 1991; Lewis & Kliewer, 1996). In addition, higher levels of negative thinking and passive adherence have been related to more visits to the emergency room (Gil et al., 1989, 1991), greater number of hospital stays (Gil et al., 1989), and increased contact with health care professionals (Gil, Thompson, Keith, Tota-Faucetter, Noll, & Kinney, 1993).

Furthermore, higher utilization of social support is associated with lower levels of state and trait anxiety and overall levels of depression (Burlew et al., 2000). However, support is the most rarely used coping strategy, with avoidance and distraction reported to be most frequently used (Zehnder, Prchal, Vollrath, & Landolt, 2006). Gil and colleagues (2001) reported that children with SCD who report using multiple cognitive and behavioral attempts to deal with pain had fewer emergency room visits and were more active during painful episodes. Children who were more passive in their approach had more health care contacts and were less physically active. Children who reported frequent negative thoughts during pain episodes had more symptoms of depression and anxiety (Gil et al., 2001). Indeed, research has demonstrated that children and adolescents with high levels of active coping attempts maintained greater levels of social, school, and home activities than those with negative thinking or passive adherence coping

styles (Gil et al., 1991, 1993, 1997). It should be noted that at least one study has reported that active coping strategies do not lead to more adaptive outcomes (Zehnder et al., 2006)

Within the coping literature, spiritual coping strategies are emerging as a more specific focus of research efforts. Although some studies suggest that spirituality can be a burden in coping with stressors, including chronic illness (Boyd-Franklin, 2003), among many African American families spirituality is viewed as a strength and resource, and many report praying as a primary coping strategy (Boyd-Franklin, 2003). Research exploring religious coping strategies shows a positive relationship between use of religious coping and use of social support and positive appraisal, suggesting a positive relationship with outcome (Britt, 1995). Although only a few studies have empirically examined religious experiences in childhood and adolescence, patients of lower socioeconomic status appear to use religious coping strategies significantly more often than those with higher SES (Landolt, Vollrath, & Ribi, 2002). Given the minority status of SCD populations, a culturally sensitive approach to treatment would necessarily include an exploration of spirtually-based coping strategies.

Family Processes. In the TSC model, family functioning is also a primary component in understanding parent and child adaptation to SCD, a perspective which is given considerable support in the empirical literature. The psychosocial adaptation of the family is positioned at the center of socioecological factors which buffer the impact of stress related to chronic illness on outcome (Drotar, 1997; Wallander & Varni, 1992). As such, it is important to explore the complex relations between a child's pain experience and subsequent pain behavior that emerge in the context of the broader family dynamics and functioning. Families construct beliefs or explanations about chronic conditions that are shaped by their larger cultural beliefs about

illness. These cognitions affect how families respond to stressful aspects of caregiving and interact with health care systems (Radcliffe, Barakat, & Boyd, 2006).

Studies regarding the functioning of families of children with SCD relative to control families have reported mixed outcomes; some families show better functioning than controls, some show more negative adaptation, while others show no differences from controls (Burlew, Evans, & Oler, 1989; Hurtig, 1994; Noll et al., 1994). Nonetheless, family functioning has been associated with coping strategies used by children with SCD and their caregivers to address SCD-related stress (Lutz et al., 2004). Children live within the context of their families, which contain rules, organizing principles, and belief systems about health, development, and illness. Therefore, the meaning of, and response to, a child's medical condition are greatly affected by the family system in which that child lives. For example, acknowledging and responding to pain may vary according to cultural and family factors (Pfefferbaum, Adams, & Aceves, 1990). Furthermore, a child's capacity to implement adaptive coping skills is influenced by the broader family environment as learning occurs through either parental modeling or reinforcement. For example, parental protective responses to pain that function as positive reinforcement, such as letting a child stay home from school or spending more time than usual with their child, have been associated with increased somatic symptoms, greater functional disability, more frequent school absences, and greater child health care utilization (Powers, Jones, & Jones, 2005). Beyond protective responses, parental minimization, defined as discounting and criticizing the child's pain as excessive, has been associated with increased somatic symptoms (Powers et al., 2005). Consistent with social learning theory, how parents respond to their child may provide cues to the child regarding his/her ability to function or elicit greater symptom reporting through positive and/or negative reinforcement.

It is important to keep in mind that "family" has become a multi-referential term in the current social environment. The word family, once used primarily to describe biological parents living together with their biological children, has given way to a broader, more diverse, and more fluid definition. Although the extended family is not unique to the African American community, the extended family remains an important cultural component due to its persistence and prevalence in the context of African American family life (Wilson, 1989). The extended family, which includes relative and fictive kin, is important not only for single-mother households, but also for two-parent families within its structure (Radcliffe et al., 2006). Although the presence of an extended family can be a source of conflict and increased stress (Murry, Bynum, Brody, Willert, & Stephens, 2001), an extended family network can also facilitate children's social adjustment, offer emotional and instrumental support, increase interaction among adults, and promote positive parenting behaviors (Taylor & Roberts, 1995; Wilson, 1989).

Family structure is often described in the SCD literature in terms of family composition and household leader. However, regardless of the type of family structure, maternal adjustment has been found to strongly influence the whole family (paternal adjustment has not been similarly studied). For example, Sharpe, Brown, Thompson, and Eckman (1994) found that families overall adaptation to SCD was heavily associated with the effectiveness of the mother's coping strategies to manage pain in the child with SCD. Conversely, mothers' use of a disengagement coping style was strongly associated with internalizing behaviors, pessimism, and negative thinking in affected children. The role of maternal coping methods has been examined in relation to children's coping methods. Klewier and Lewis (1995) found less parental use of cognitive restructuring and more parental use of active coping strategies predicted children's use of avoidance coping. Coping strategies of parents of children with SCD also predict a significant

portion of the variance in the child's adjustment to SCD beyond that accounted for by the child's age and frequency of painful episodes (Gil et al., 1991). In some cases, mothers of adolescents with functional limitations because of chronic pain have been found to interact with their children in ways that discourage adaptive coping (Dunn-Grier, McGrath, Rourke, Latter, & D'Astous, 1986).

Families of children and adolescents with SCD have a high number of characteristics that may influence their responses to SCD (Radcliffe et al., 2006), including having a higher likelihood of single-parent structure, relying on extended family support and flexible family roles, stress related to low socioeconomic status (i.e., perceived poverty, lack of employment opportunities, reduced access to health care, and living in stressful urban environments), and factors associated with ethnic minority status. Although these characteristics may be perceived as risk factors, and indeed poverty has been linked to poorer psychological outcomes particularly for African American families in which highly stressed parents engage in less-effective parenting practices (Radcliffe et al., 2006), they also bring a set of strengths, strategies, values, and capabilities that can support resilience (Boyd-Franklin, 2003). Such significant strengths of African American families may include a strong work orientation, strong and consistent extended kin networks and adaptability of family roles, high achievement orientation, resourcefulness, and strong religious orientation (Radcliffe et al., 2006).

Cognitive-Behavioral Approach to Pain Management

Individual Approach. Although medication has been by far the treatment primarily utilized for SCD (and even though medication has limited efficacy for pain management), research has strongly supported use of pain management strategies that incorporate

psychological intervention for patients with SCD. Feelings of helplessness, loss of control, depression, and anxiety symptoms during painful episodes can exacerbate pain and influence the pain presentation of SCD (Pallister, 1992). A small number of studies have demonstrated that cognitive-behavioral therapy (CBT) procedures for general pain management (which include techniques such as guided imagery, changing maladaptive thinking, distraction, progressive muscle relaxation, and breathing exercises), result in improvements in children's recurrent and chronic pain symptoms (Powers et al., 2005). As a result, CBT is the standard psychological intervention for general pain (Morley, Eccleston, & Williams, 1999) guided by the premise that developing and implementing a variety of coping strategies for dealing with pain will improve adjustment (Haythornthwaite, Menefee, Heinberg, & Clark, 1998). CBT interventions which specifically target adaptational responses to pain have demonstrated not only pain reductions but also increases in adaptive coping responses, self-efficacy, physical functioning, and decreases in maladaptive cognitions (Gil et al., 1997; Turner & Jensen, 1993). It is noteworthy that there are no existing evidence-based CBT interventions which target pain management for pediatric patients with SCD (see Chen et al., 2004, for a review of related interventions).

However, a handful of studies have used CBT strategies for SCD pediatric populations, with varying degrees of success. In a randomized study of children with SCD, a brief cognitive-behavioral one-session (plus review session) intervention that taught deep breathing, imagery, and self-statements (Gil et al., 1997) was tested. Although after the intervention, children did not differ from the control group on reported pain, within group analyses revealed that the use of active coping strategies on high pain days was associated with decreases in negative thinking, fewer health care contacts, less reduction in daily activities, and fewer school absences (Gil et

al., 2001). These results suggest that brief coping skills training may result in lower levels of negative thinking and reported pain during low levels of pain stimulation.

Intervention studies for adolescents with SCD not directly targeting pain have found some related improvements in symptoms. An 8-week group psychoeducation intervention with adolescents with SCD found some improvement in the ability to manage pain (Baskin et al., 1998). Also, in a study investigating the efficacy of a support group intervention for adolescents with SCD, patient well-being was positively related to satisfaction with the support group and to absence of SCD complications, including pain (Telfair & Gardner, 1999). Although both these studies demonstrated that adolescents respond favorably to psychosocial intervention designed specifically for this age group, neither study focused on pain management as a direct target of intervention for adolescents with SCD.

Family Approach. Powers and colleagues assessed the effectiveness of family-based CBT intensive pain management for a small sample of children with SCD, demonstrating improvements in child coping and daily functioning (Powers et al., 2002). Empowerment of the family system was viewed as the key feature to therapeutic change for these families (Boyd-Franklin, 2003). Empirical evidence shows that involving parents in the treatment of pediatric chronic pain is important in maintaining treatment gains related to the effects of modeling and reinforcement over time (Chambers, 2003).

Powers Mitchell, Graumlich, Byars, and Kalinyak (2002) investigated the feasibility and outcome of a CBT program for children (ages 9-12 years) with SCD and their primary caregivers. Parents participated in parent education/skills training and played an active role in helping their children complete pain diaries, take medications and use coping skills optimally. Children participated in a 6-session intensive cognitive behavioral program that integrated both

pharmacological and nonpharmacological strategies (i.e., education about SCD and pain medications, deep-breathing, self-monitoring, progressive muscle relaxation, distraction, positive self-talk, and physiological techniques) for pain management. Although limited in sample size (n=3), results indicated that participants demonstrated a decrease in negative thinking, and one participant showed an increase in active coping attempts.

Cultural Sensitivity. Kaslow and colleagues (Kaslow et al., 2000; Kaslow & Brown, 1995) were among the first to argue that to be most effective, interventions with children and adolescents with SCD must be culturally sensitive. They developed and tested a six-session intervention for children and adolescents with SCD ages 7 to 16 and their families that included culturally sensitive components; these included involvement of family and focus on family strengths, collaborative nature of the intervention, training of the research team on cultural sensitivity with African Americans, and diversity of the research team. In addition, the treatment was individualized for each child and family in terms of their unique needs and competencies. This intervention resulted in improved disease knowledge of parents and children, but no differences in distress or family functioning were found post-treatment. Unfortunately, the study did not assess changes in pain.

Overall then, research clearly shows the positive impact of skills-focused interventions targeting behavioral and cognitive coping strategies for the individual with SCD. Cognitive-behavioral techniques show some success in leading to pain reduction and increases in positive coping in SCD, however, a number of barriers stand in the way of conferring evidence-based status to these interventions. Maintenance of treatment effects has not been documented (Gil et al., 1997), as SCD intervention follow-ups have not been reported beyond 6- months post-treatment. Furthermore, sample sizes have been small, although this is not surprising given the

difficulty in engaging African Americans in intervention research (Green, Patridge, Fouad, Kohler, Crayton, & Alexander, 2000). In addition to individual approaches, research has identified the importance of family-based intervention. From an ecological standpoint, individual skills, and a child's ability to implement these skills, are heavily influenced by the family context, as parents serve as models for coping, and provide encouragement and reinforcement of pediatric pain behaviors. In other words, the adaptational processes of the pediatric patient (and CBT skills targeted during individual interventions) are closely interrelated with parent and family processes, and may vary in their success as a direct function of the family context. Finally, although CBT interventions have yielded some positive outcomes for SCD patients, given the unique characteristics of the SCD population it is likely that greater benefits will come with modifications to allow more flexibility with the individual patient's unique presentation needs, developmental and gender differences, family resources and limitations, and preferences for varying coping strategies (i.e., spiritually-based strategies), etc. (Schwartz, Radcliffe, & Barakat, 2007). Given these considerations, the present study will provide a family-based pediatric cognitive-behavioral pain intervention that will target individual cognitive processes, coping strategies, and family processes.

Present Study

The present study proposes to assess the feasibility of a treatment targeting both pain and psychosocial symptoms for SCD through skills training, family involvement, and cognitive-behavioral techniques. Guided by the transactional stress and coping model, the present study will modify aspects of an existing, *cognitive-behavioral family intervention (CBFT)* for pain management with children and adolescents experiencing Recurrent Abdominal Pain (RAP) for

the treatment of children and adolescents with SCD (Robins, Smith, Glutting, & Bishop, 2005), which will be described in detail later.

Although this pain management intervention has only targeted children, adolescents, and families with RAP, similarities can be noted between RAP and SCD, primarily in the occurrence of pain symptoms, psychosocial and psychological concerns, limited functionality, less satisfactory quality of life, and disruptions to the family (Hocking & Lochman, 2005; Merlijn et al., 2005). This CBFT intervention is brief and has demonstrated significant reductions, both short- and long-term (approximately 1-year), of pain symptoms, school absences, health-care contacts, as well as improvements in daily functioning, mood, and coping strategies utilized (Robins et al., 2005; Sanders, Sheperd, Cleghorn, & Woolford, 1994). Given these similarities, it is reasonable to expect that an intervention shown to be effective for RAP could be successfully modified to target the SCD population. The primary aims of the present study were as follows:

1. Extend the Robins et al., 2005 CBFT intervention for RAP to the SCD pediatric population using an individualized and culturally-sensitive approach.
2. Evaluate effectiveness of the CBFT intervention for reducing pain symptoms, improving HRQoL, decreasing functional limitations, and improvements in psychological adjustment.

Hypotheses

As stated previously, this study will provide a CBFT intervention that targets pain symptoms and psychosocial functioning in children and adolescents with SCD. Four main outcome areas were assessed during the course of the CBFT intervention in order to quantify treatment change. First, *reduction in pain symptoms* were evaluated based upon measured

change in frequency and intensity of pain symptoms. Second, *improvement in HRQoL* was measured by using a standard assessment of life satisfaction. Third, *decreases in functional limitation* was assessed using a general functional disability inventory and by monitoring health care contacts. Fourth, *improvements in psychological adjustment* were assessed via changes in anxiety, depression, and coping strategies utilized. Using a single case study design with nonconcurrent multiple baseline methodology (Carr, 2005), the following specific hypotheses were tested:

Hypothesis 1: Baseline Changes. During the baseline phase (prior to intervention) it was expected that the intensity of pain symptoms, functionality, and anxiety would be relatively stable. Ratings of pain symptoms, functionality, and anxiety (as reported in daily diaries) would remain unchanged during baseline.

Hypothesis 2: Reduced Symptoms of Pain. It was expected that ratings of pain symptoms would change as a function of the therapeutic intervention.

a. Compared to baseline, a decrease in the intensity of pain symptoms (as reported in daily diaries) was expected during the treatment phase.
b. Compared to baseline, a decrease in pain symptom intensity (as measured on the PPQ) was expected at post-treatment (2-, 4-, and 6-month post-treatment).

Hypothesis 3: Decreases in Functional Limitations. It was expected that ratings of functional limitation would change as a function of the therapeutic intervention.

a. Compared to baseline, fewer functional limitations (as reported in daily diaries) were expected during the treatment phase.
b. Compared to baseline, fewer functional limitations (FDI) were expected at post-treatment (2-, 4-, and 6-month post-treatment).

Hypothesis 4: Improvement in HRQoL. It was expected that ratings of HRQoL (PedsQL) would change as a function of the therapeutic intervention. Life satisfaction and well-being were expected to improve from baseline to post-treatment (2-, 4-, and 6-month post-treatment).

Hypothesis 5: Improvements in Psychological Adjustment. It was expected that ratings of psychological adjustment would change as a function of the therapeutic intervention.

a. Compared to baseline, fewer symptoms of anxiety (as reported in daily diaries) were expected during the treatment phase.
b. Compared to baseline, fewer symptoms of anxiety (RCMAS) and depression (CDI) were expected at post-treatment (2-, 4-, and 6-month post-treatment).
c. Compared to baseline, internalizing and externalizing symptoms (CBCL were expected at post-treatment (2-, 4-, and 6-month post-treatment).
d. Compared to baseline, more frequent use of positive coping strategies (CSQ-R) were expected at post-treatment (2-, 4-, and 6-month post-treatment).

Methods

Design

A nonconcurrent multiple baseline design was implemented with participants beginning treatment after baselines of two, three, or four weeks. This design is essentially a series of A-B replications in which the length of the baseline phase differs in a pre-arranged but random way (Carr, 2005). The experimental control demonstrated by the multiple baseline design across participants can be described using three elements of single-case design logic (Johnston & Pennypacker, 1993). First, repeated measures can establish the prediction of a baseline's data

path into the subsequent treatment phase, allowing the detection of a difference between the actual data path in treatment and the path predicted from baseline (Petermann & Muller, 2001). Second, the effects of the independent variable are verified by demonstrating that the intervention changed the participant's behavior (Petermann & Muller, 2001). Finally, the effects of the independent variable are replicated across different participants (Petermann & Muller, 2001). This design helps to ensure that the symptoms of participants are not transient and changes are not associated with the seeking of treatment or the assessment process.

Participants

Participants in this study were eight youth (4 males, 4 females) with SCD and their parents (or designated caretakers). The children's ages ranged from 8 to 15 years (mean age = 11.88, *SD* = 2.42). All participants were African American. Of the eight children and adolescents, 3 (37.5%) lived in two-parent households, whereas 5 (62.5%) lived in single-parent households. For the reporting parent, the mean age was 40.86 (*SD* = 8.47, range = 30 – 53), and the mean education level was 14.5 years (*SD* = 1.81, range = high school graduate to college graduate). Socio-economic status was the parent's educational level plus their occupation (see Hollingshead (1975) Index of Social Status). Socio-economic status was organized as low or middle, coded as 0 and 1 respectively. Participant demographics are summarized in Table 1.

All participants were diagnosed as having SCD by standard laboratory methods, including hemoglobin electrophoresis. Participants were categorized into two phenotypes: sickle cell anemia (Hb SS), or hemoglobin SCD (Hb SC). Although there is considerable variability across the phenotypes, Hb SS is usually the more clinically severe type (Gil et al., 2000). Five participants were currently receiving hydroxyurea treatment, which is considered the usual and

customary medical treatment for severe pain in SCD. Exclusion criteria included: (a) currently receiving cognitive or behavioral therapy, (b) estimated IQ less than 80, or (c) current homicidal ideation. No participants were ineligible for the study. Participant flow is shown in Figure 2. Eleven children and adolescents were consented for the current study. Three youth and their families declined to enter treatment. Of the eight children who began treatment, none dropped out of the study prior to completion of the intervention. Two participants did not complete follow-up measures.

Participant 1, a 14-year-old African American male, lived with his biological father, step-mother, and younger step- and half-siblings. The family was of middle socioeconomic status. He has a history of acute chest syndrome, a respiratory complication, and treatment of blood transfusions. The participant was taking hydroxyurea and penicillin daily throughout treatment. As part of the typical pain management protocol, the participant was prescribed opioids (e.g., Tylenol with codeine) on an as needed basis. Prior to treatment, the participant was hospitalized three times in 2009 for pain crisis, each hospitalization lasting approximately ten days.

Participant 2, a 13-year-old African American female, lived with her biological parents and older biological siblings. The family was of middle socioeconomic status. The participant was taking hydroxyurea throughout treatment. As part of the typical pain management protocol, the participant was prescribed opioid (e.g., Tylenol with codeine) and nonsteroidal anti-inflammatory (e.g., ibuprofen) medications on an as needed basis. Prior to treatment, this participant had not been hospitalized for a pain crisis since 2003.

Participant 3, an 8-year-old African American male, lived with his biological parents and older biological brother. The family was of upper middle class. The participant was prescribed

Tylenol with codeine on an as needed basis throughout treatment. Prior to treatment, this participant had not been hospitalized for a pain crisis since 2006.

Participant 4, a 13-year-old African American male, and Participant 5, 9-year-old female, were half-siblings residing with their maternal grandmother. Their biological mother passed away in 2008 from complications of her SCD; participants were not in contact with their biological father. The family was of lower middle socioeconomic status. Participant 4 was taking hydroxyurea and folic acid daily throughout treatment. Prior to treatment, this participant had not been hospitalized for a pain crisis since 2005. This participant also met diagnostic criteria for stuttering and PICA. At baseline, he endorsed suicidal ideation (i.e., "think about killing myself but I would not do it"). Participant 5 was taking hydroxyurea and folic acid daily throughout treatment. She has a history of splenic sequestration, a gastrointestinal complication. Prior to treatment, this participant was hospitalized twice in 2009, only once for a pain crisis.

Participant 6, a 15-year-old African American female, lived with her biological mother. Her biological father passed away in 2005. The family was of middle socioeconomic status. She has a history of splenic sequestration, a gastrointestinal complication, and acute chest syndrome, a respiratory complication. The participant was taking hydroxyurea throughout treatment. As part of the typical pain management protocol, the participant was prescribed nonsteroidal anti-inflammatory (e.g., ibuprofen) on an as needed basis. Also at baseline, she endorsed that she "thinks about killing myself but I would not do it." Prior to treatment, this participant was hospitalized in January 2010 for gallbladder and kidney complications, requiring a blood transfusion at the time.

Participant 7, a 12-year-old African American male, lived with his biological mother, and did not have contact with his biological father. He was home-schooled. The family was of

middle socioeconomic status. This participant was previously seen by a psychiatrist for PICA. This participant was prescribed Tylenol with codeine on an as needed basis. Prior to treatment, he was last hospitalized in 2002 which required a blood transfusion.

Participant 8, an 11-year-old African American female, lived with her biological mother and younger biological sibling. She had no contact with her biological father. The family was of middle socioeconomic status. During baseline and the majority of treatment sessions the participant was not taking any medication; she began hydroxyurea after the fourth treatment session (two weeks prior to treatment completion). Prior to treatment, she was hospitalized twice in 2009 for fever, pain crisis, and acute chest syndrome.

Measures

Once participants had been selected based on the above stated inclusion and exclusion criteria, children, adolescents, and their families were administered several baseline assessment measures. Youth and their parents completed several questionnaires at each assessment time point (baseline, post-treatment, 2-, 4-, and 6-months post-treatment). These measures assessed symptoms of pain, HRQoL, functionality, coping strategies (general and religious) and psychological adjustment outcomes. A brief description of each measure and associated psychometric properties is presented below.

Varni/Thompson Pediatric Pain Questionnaire (*PPQ*; Varni, Thompson, & Hanson, 1987). (*Parent and Child Version*). The PPQ is a self- and caregiver report measure that combines several modalities to assess different aspects of pain. The PPQ addresses evaluative, sensory, and affective components of pain (pain descriptors as identified by child or parent) and pain location (body outline of pain sites). The visual analogue scales (VAS) are used to assess

the present and worst by placing a mark on a horizontal line anchored from "no pain" to "severe pain." The PPQ has reported a test-retest reliability of .61-.90 and convergent validity of .27-.68 with disease status, and .06-.45 with psychological functioning (Varni et al., 1987).

Daily Pain Diary (Hunfeld et al., 2001; Shapiro et al., 1990). (*Parent and Child Version*). (see Appendix A). Throughout treatment a daily pain diary will be used to record frequency and intensity of daily pain episodes using the VAS ("no pain" to "worst pain"). The VAS has demonstrated a concurrent validity of .61-.90 and test-retest reliability of .41-.58 (Hunfeld et al., 2001, 2002). In addition, the location of pain will be indicated using a body diagram, activity reduction will be noted, treatment adherence, and psychological and nonpharmacologic pain-relieving techniques were also recorded.

Coping Strategies Questionnaire-Revised Sickle Cell Disease Version for Children (*CSQ*; Gil et al., 1989). The CSQ consists of 80 items rated by children and adolescents that measures how often participants use cognitive, behavioral, and physiological coping strategies during sickle cell disease-related pain. Respondents rate the frequency of use of strategies on a 7-point Likert-type scale ranging from 0 "never use this strategy" to 6 "always use this strategy". The CSQ produces 3 factor scores: Coping Attempts, which measures "active" coping strategies such as talking with someone, going on despite pain, play outside; Negative Thinking, which includes behaviors such as pessimistic thinking and worry; and Passive Adherence, which includes behaviors such as praying, resting, and taking medications. Internal consistency is shown for all 3 factors scales with alpha reliabilities ranging from .55 to .72 for Coping Attempts, .67 to .70 for Negative Thinking, and .66 to .89 for Passive Adherence, respectively (Gil et al., 1991). Internal consistency for the current sample, as measured by Cronbach's alpha, ranged from .85 to

.94 for Coping Attempts, from .74 to .94 for Negative Thinking, and from .74 to .90 for Passive Adherence across the various assessment time points.

Child Spiritual Coping Survey (*CSCS;* Boeving, 2003). The CSCS is a 22-item measure of religious (9 items) and existential (13 items) coping strategies. For religious and existential coping, a range of possible total scores for both frequency and efficacy is 0-36 and 0-52 respectively. The administration of the measure involves a prompt by the interviewer to "think about when you are feeling down or afraid about being sick." This child is then asked to rate each coping item based upon frequency of use and effectiveness of techniques to reduce distress. Response options range from 0 ("I never do this"/ "This never helps") to 4 ("I always do this"/ "This always help"). Higher scores equate to more frequent use and effectiveness of such strategies. Internal consistency reliability estimates, for both the frequency and efficacy items, on the religious coping subscale were high .93 and .92, while the existential coping subscale was slightly lower, although acceptable, .86 and .87 respectively (Boeving, 2003). In the current sample, Cronbach alpha's were comparable to the previous study for both frequency and efficacy items. On the religious coping subscales, frequency ranged from .86 to .97, and efficiency ranged from .95 to .97 from baseline through 6-months post-treatment follow-up. Comparatively, on the existential coping subscales, frequency ranged from .89 to .96, and efficacy ranged from .86 to .96.

Pediatric Quality of Life Inventory (*PedsQL*; Varni, Seid, & Rode 1999). (*Parent and Child Version*). The PedsQL is a measure of health-related quality of life administered to pediatric populations. The measure consists of 23 items that comprise four scales: physical, emotional, social, and school-based functioning. A total score is obtained by summing all items over the number of items answered on all scales. The instructions ask how much of a problem

each item has been during the past month. A 5-point response scale is used across child self-report for ages 8-18 and parent proxy-report (0 = never a problem; 1 = almost never a problem; 2 = sometimes a problem; 3 = often a problem; 4 = almost always a problem). Items are reverse-scored and linearly transformed to a 0-100 scale (0 = 100, 1 = 75, 2 = 50, 3 = 25, 4 = 0), so that higher scores indicate better HRQoL. The PedsQL has demonstrated an internal consistency of .68 to .90, and a cross informant correlation coefficient ranging from .36 to .50; however, no test-retest reliability has been report (Varni, Seid, & Rode 1999). The internal reliability (Cronbach's alpha) of the total score in the present study ranged from 0.82 to .95 for youth report across the five assessment points, and ranged from .88 to .96 for parent report from baseline to 6-month post-treatment follow-up.

Functional Disability Inventory (*FDI*; Walker & Greene, 1991) (*Parent and Child Versions*). The FDI assesses physical and psychosocial limitations (e.g., "walking up the stairs," "doing something with a friend"), as reported by children, adolescents, and their caregivers, during the past two weeks on a 5-point Likert scale (0= "no trouble" to 4="impossible"). Scores range from 0 to 60 with higher scores indicating more disability. The FDI has demonstrated an internal consistency of .85-.92, test-retest reliability of .74 for 2-weeks, and .48 for 3 months, and evidence of validity was examined by correlations with school absence (r =.44, p<.001) and somatic symptoms (r =.45, p<.001) (Walker & Greene, 1991). The internal reliability of the total score in the present study ranged from .81 to .90 for youth report across the five time points, and ranged from .91 to .96 for parent report throughout the study.

Number of health care contacts (hospitalization, emergency room visits) will be obtained via parent-report and from the hospital electronic databases for the 6-12-month period prior to

and following study entry, as a measure of health-related outcomes and functional impact (e.g., impact on child's daily activities).

Children's Depression Inventory (*CDI*; Kovacs & Beck, 1977). The CDI is a 27-item self-report instrument scored on a 3-point scale. Each item is scored according to the presence of the specified symptoms (0=absence of a symptom, 2=severe symptom presence). The sum of the item scores result in the total score, which can range from 0 to 54. Higher scores indicate greater depressive symptomatology. Normative data indicate that a combined male and female sample mean is 9.09 with a standard deviation of 7.04. The upper 10% of the distribution is indicated by a cutoff score of 19. In a sample of normal children ages 8-16, the CDI has shown a range of internal consistency coefficients between .83-.89. However, test-retest reliability has been variable across populations and testing intervals. Validity studies indicate that that the CDI correlates highly with measures of self-concept and distinguishes emotional distress from normal school-age children. Internal consistency for the current sample ranged from modest, 0.70, to high .94 across the five assessment time points.

Revised Children's Manifest Anxiety Scale (*RCMAS*; Reynolds & Richmond, 1985). The RCMAS is a 37-item (yes/no) self-report inventory that assesses the level and nature of anxiety in children and adolescent in four areas: physiological anxiety, worry, social concerns, total anxiety. The RCMAS can be used with 6- to 18-year-old children and adolescents. It has been recommended that an overall cut-off point of 19 to 28 be used to identify children experiencing clinically significant levels of anxiety. The RCMAS has been shown to have an internal consistency reliability greater than 0.80 and good convergent reliability (Reynolds & Richmond, 1978). Internal consistency for the current sample ranged from .82 to .97 from baseline through 6-month post-treatment follow-up.

Demographic Information about the child, parents, and family situation will be gathered via a two-page form completed by parents. Child information included the child's age, race, gender, grade in school, and basic information regarding illness (e.g., number of emergency room visits and overnight hospital stays in the past twelve months). Parent information included age, marital status, education level, occupation, income, and social support.

Adult Responses to Child and Adolescent Symptoms (*ARCS*; Van Slyke & Walker, 2006). The ARCS is a 33-item measure of parent responses to their child's pain across three scales: protectiveness, minimization of pain, and encouraging/monitoring responses. Parents rate the helpfulness of each source on a 5-point Likert scale (0="never" to 4="always") according to how often they use each strategy, and subscale scores are computed by calculating the mean ratings for items on each subscale. Higher scores equate to more frequent use of a particular response. Items on the Protect scale refer to protective parent behavior such as giving the child special attention and limiting the child's normal activities and responsibilities. Items on the Minimize scale discount and criticize the child's pain as excessive. Items on the Distract and Monitoring scale refer to assessing the child's symptoms (e.g., asking questions and checking on the child). Internal consistency is shown for all 3 scales with alpha reliabilities of .87, .67, and .79 for Protect, Minimize, and Encourage/Monitor, respectively (Van Slyke & Walker, 2006). Internal consistency for all 3 scales in the present study ranged from .90 to .98 for Protect, .46 to .55 for Minimize, and .78 to .97 for Distract and Monitor across baseline throughout 6-month post-treatment follow-up.

Pediatric Inventory for Parents (PIP; Streisand, Braniecki, Tercyak, & Kazak, 2001). The PIP is a 42-item parent-report measure aimed specifically at measuring the nature of disease related parenting stress. Parents rate a list of general, medically related situations and thoughts

considered stressful to parents of children with an illness along a 5-point Likert scale (1 = "Not at all," 5 = "Extremely") as to both the item's frequency over the last week and level of difficulty associated with it. The following disease-related parent-child domains include communication, emotional functioning, medical care, and role function. Frequency and difficulty scores are summed separately for each of the four domain scales. These scale scores are then added together to form an overall total frequency score (PIP-F) and total difficulty score (PIP-D): higher scores indicate greater frequency and difficulty. The PIP has demonstrated high internal consistency reliability with alpha reliability ranging from .80 to .96 (Streisand et al., 2001). PIP scores were significantly correlated with a measure of state anxiety and also with parenting stress, demonstrating construct validity in a sample of pediatric cancer and juvenile diabetes samples (Streisand et al., 2001). For the current study, a high Cronbach's alpha was shown for both the total frequency and total difficulty scores, which ranged from .90 to .97, to .86 to .96 respectively.

Child Behavior Checklist (*CBCL*; Achenbach & Edelbrock, 1983). The CBCL is a parent-report checklist of children's behavior problems (internalizing, externalizing and total problems). Parents rate each item 0=not true, 1=sometimes true, or 2=often true of their child's behavior. This instrument can be used with 4- to 18-year-old children and adolescents. T-scores can be derived for both internalizing and externalizing problems as well as total behavior problems. T-scores of 60 to 63 (84^{th} to 90^{th} percentile), and 64 or above (greater than the 90^{th} percentile) for total behavior problems, internalizing and externalizing problems are considered to be in the borderline to clinically significant range of functioning. The CBCL is widely used demonstrating test-retest reliabilities ranging from 0.95 to 1.00, inter-rater reliabilities ranging

from 0.93 to 0.96, and internal consistencies ranging from 0.78 to 0.97 (Achenbach & Edelbrock, 1983).

The following table illustrates the administration schedule of each instrument:

Baseline	Treatment (Sessions 1-5)	Post-Treatment	2-month Post-Treatment	4-month Post-Treatment	6-month Post-Treatment
Parent:					
Demographics PPQ PedsQL FDI ARCS PIP CBCL				PPQ PedsQL FDI ARCS PIP CBCL	
Child:					
PPQ CSQ CSCS PedsQL FDI CDI RCMAS				PPQ CSQ CSCS PedsQL FDI CDI RCMAS	
Parent & Child:					
Daily Pain Diaries					

Procedure

Participating youth were recruited through the Pediatric Sickle Cell Program at Brenner Children's Hospital (affiliated with Wake Forest University Baptist Medical Center in Winston-Salem, North Carolina). Eligible families (i.e., having a child between the ages of 8 and 15 diagnosed with SCD) were first contacted by mail to explain the study. Families were invited to participate by contacting the research coordinator. Interested families were then scheduled for a

comprehensive baseline assessment to determine eligibility for the treatment protocol. Written informed consent was obtained from one parent or caregiver and assent was also obtained from the child or adolescent (see Appendix B).

Participants were randomly assigned to baseline phases lasting two, three, or four weeks (one participant in the two week condition, four participants in the three week condition, and three participants in the four week condition). During each week of baseline, youth and their parents completed daily diaries to establish pain trends prior to intervention. Following the baseline period, the intervention phase began (outlined below). During the intervention phase, completion of daily pain diaries continued (i.e., repeated observations measurement of the dependent variable; Harvey, May, & Kennedy, 2004).

The CBFT intervention was scheduled bi-monthly for a total of five 50-minute sessions. Each participant was paid $10 for the baseline assessment and for each additional treatment session in which they participated (for a possible total of $60 for the assessment and treatment sessions). Participations were also compensated for completion of post-treatment assessment measures, $20 at 2-, 4-, and 6-months post-intervention assessment. Each participant had the potential to receive $120 in compensation for their time and participation.

Treatment

The five session CBFT intervention targeted the use of specific problem-solving skills, use of effective cognitive and behavioral coping strategies, relaxation, and goal setting. The CBFT intervention consists of 4 pivotal components for both parent and child: (1) education about the credibility of CBT to establish an active collaboration between child, family, and therapist, (2) skills acquisition which will allow the child to achieve a sense of control over their

pain, (3) cognitive and behavioral rehearsal to allow child and parent to actively learn new behaviors and cognitions to better manage pain (skills which may include learning self-regulation skills, progressive muscle relaxation, and goal setting), and (4) generalization and maintenance of skills to facilitate skill retention and avoid increases in pain following treatment completion (Bradley, 1996). Parents and children were seen conjointly for three of the five sessions, and there was a review of the goals and activities with both parent and child before and after each session. Parents were encouraged throughout to become proficient in the same skills as their children. Homework was provided following all sessions to allow further practice of skills. Flexibility in scheduling was provided to all families, resulting in the majority of sessions taking place in the evenings and on the weekends. Other culturally sensitive modifications are provided in the table at the end of this section. Specific session goals are detailed below.

Session 1: The objectives for both parent and child were to (a) develop an understanding of the child's pain, (b) increase repertoire of pain management techniques, and (c) increase understanding of the connection between stress and pain perception. Assessment was again, conducted through a detailed clinical interview with the child and parent. The frequency, duration, location, intensity, antecedents, and consequences of pain were assessed to thoroughly characterize the child's pain. Second, the educational component presented a credible rational for the cognitive-behavioral intervention designed to elicit the active collaboration of pediatric patients and families with the therapist. Patients were actively encouraged to adopt the belief that he or she [they] can learn the skills necessary to better cope with pain and other illness-related problems. Modeling and practice of pain management techniques were introduced (i.e., breathing, imagery, and relaxation techniques) and the connection between stress and pain was discussed.

Sessions 2 and 3: These sessions were with the child and adolescent only, with the primary aim of helping pediatric patients achieve control over their pain. Specific objectives were as follows: (a) increase repertoire of pain management techniques, (b) encourage the child to "take control" of pain, and (c) learn to challenge negative predictions, learn positive self-statements, and identify the associated impact on pain. This component was intended to help patients engage actively in the process of learning new behaviors and cognitions so they can better manage pain and other pain-related problems. Children practiced their new pain management behaviors and cognitions and applied these behaviors and cognitions to their home and school environments. Children also worked to alter their negative perceptions regarding their ability to manage pain and the psychological consequences of pain. To meet these objectives, children reviewed daily pain diaries, focusing on the antecedents and consequences of their pain episodes. "Self-talk" was introduced, along with the use of positive self-statements to challenge negative predictions. The concepts of snowballing, catastrophizing, and distraction techniques were also described.

Session 4: This session involved both the child and parent, with the objective of increasing their "partnership" in the active management of pain. This was achieved by instructing parents to reframe their role from "protector" to "coach." Parents were encouraged to minimize their discussion of pain, reinforce their child's behaviors that are incompatible with being sick, and limit the child's secondary gains from their sick behaviors. Discussion also included the parent's own coping strategies and the role this may play in their child's pain experience.

Session 5: Progress was assessed with both the child and parent and treatment gains were reinforced. The goals of this generalization and maintenance session were to help children retain

their learned skills and avoid increases in pain post-treatment, as well as to encourage parents in the implementation of their more adaptive responses. Preparation for continued coping was addressed by reviewing relaxation skills, cognitive techniques, and distraction tools. Post-intervention measures were completed. Families were reminded by mail and by phone call of follow-up assessments.

Culturally Sensitive Modifications

	Culturally Sensitive Component	Present Study Examples
Session 1	• Awareness of stigma of mental health problems and mistrust of medical research	• Medical team (which consisted of Hispanic physician and African American nurse practitioner and social worker) assisted with recruitment and provided support for the intervention • Objectives of study were clearly reviewed during informed consent/assent procedures • Intervention framed as a supplement to existing medical care, i.e., as an additional way to manage disease • Acknowledged and validated various concerns related to research and limited resources • Weekly phone contact, reminders for sessions
	• Culturally sensitive content	• Language of "I" statements were flexible and chosen by adolescent (e.g., included content from the "Bernie Mac" Show) • Participants chose imagery that was personally exciting and active (e.g., sporting events, familiar setting, scenes from favorite television shows) • Adolescents identified relaxing music to listen to • Inclusion of culturally sensitive measures (e.g., African American figures)

	• Family-based	• Family members attended sessions and worked with youth to implement pain management strategies
Session 2 & 3	• Emphasized empowerment	• Skills training to enable youth and family to take control of managing pain (i.e., encouraged self-advocacy) • Self-statements included ethnic/racial pride and faith related content
Session 4	• Family-based	• Family members attended session and worked with youth to implement pain management strategies • Collaborate to develop realistic and feasible pain coping strategies
Session 5	• Emphasized empowerment	• Assessed for and reinforced adaptive coping skills and resources for dealing with SCD and related pain

Therapist. All participants were treated by the current author who was a masters-level graduate clinician enrolled in an APA approved doctoral program in clinical psychology. The clinician was supervised by a licensed clinical psychologist and the protocol was implemented in a clinically and culturally sensitive and flexible manner.

Treatment Adherence. Two independent trained coders rated audio transcriptions of all sessions for each participant to assess the degree of treatment adherence. The transcriptions were rated based on adherence to identified elements of the intervention that were targeted during each session. The Adherence Checklist consisted of 8-16 "yes/no" elements per session (see Appendix C). From these ratings, session adherence scores and a total adherence score across all five sessions were calculated (interrater reliability as 100%). Raters indicated mean session and total adherence scores of 100% for the therapist, which reflects a high degree of adherence to the treatment protocol.

Results

One of the more common and most straightforward methods for evaluating multiple baseline studies is through visual inspection and noting whether gains were made during the intervention (Carr, 2005). Differences between baseline and the treatment phase were evaluated for differences in the *mean* (e.g., average frequency of symptoms), *level* (e.g., whether the change was stable), changes in the *trend*, or slope (either increase or decrease) of the data, and the *latency* of change (how quickly treatment gains were observed) (Kazdin, 1998). Consistent with single case methodology, each outcome is summarized below to examine changes in means, levels or trends, and latency of change across baseline and treatment for each participant. Results presented below are organized according to the outcome of interest. A description of results from daily diaries for symptoms of pain and functional limitations is presented first, followed by HRQoL and psychological adjustment outcomes.

Overall, the present study demonstrated support for some, but not all of the hypotheses. Results are briefly summarized first and then presented in detail below. *Hypothesis 1*. During the baseline phase, five of the eight participants (2, 3, 4, 5, and 7) endorsed stable pain intensity ratings as reported in daily diaries; ratings of functionality were stable for five participants (3, 4, 5, 6, and 7); and symptoms of anxiety were stable for five participants (2, 4, 5, 6, and 8). Thus, stability in pain and psychological adjustment was established for some, but perhaps not all participants. *Hypothesis 2.* Compared to baseline, a mean decrease in pain intensity (as reported in daily dairies) was observed for five participants (2, 3, 4, 6, and 7) during treatment. However, significant differences were not observed in pain intensity from baseline to post-treatment or follow-up (as measured on the PPQ). *Hypothesis 3.* Compared to baseline, fewer functional limitations were reported by four of the eight participants (2, 3, 4, and 8) during treatment

(reported in daily diaries). In contrast, no significant changes in functionality were observed from baseline to follow-up (as measured on the FDI). *Hypothesis 4.* Significant improvements in HRQoL from baseline to post-treatment were demonstrated by all participants and these improvements were maintained at the 2, 4, and 6-month post-treatment follow-ups (as measured on the PedsQL). *Hypothesis 5.* Compared to baseline, decreases in symptoms of anxiety (as reported on the daily diaries) were observed for two participants (1 and 3) during treatment. Anxiety symptoms (as measured on the RCMAS) were decreased for all participants at 2, 4, and 6-months post-treatment. In addition, as compared to baseline, youth reported significant decreases in depressive symptomatology (as measured on the CDI). These effects were maintained at the 2-, 4-, and 6-month follow-up. Compared to baseline, parents reported significant decreases in internalizing, externalizing, and total behavior problems (as measured on the CBCL), which were maintained across 2, 4, and 6-months post-treatment. Lastly, while not statistically significant, changes in general coping strategies were observed, including increases in use of active coping strategies at post-treatment follow-ups (with a large effect size). Similarly, a decrease in passive adherence coping strategies was observed at post-treatment (with a large effect size). Some decreases in negative thinking were reported at post-treatment (with small effect size), but not maintained at follow-up.

Weekly Pain Symptoms

Initially, diary responses from youth and their parents were compared to determine the degree of agreement. Of the eight participants, six parent-youth dyads had significantly consistent reports of pain ratings, with correlations ranging from .3 to .869 (see Table 2 for correlations). The two cases of discrepant ratings occurred in the same family (Participants 4 and 5), with the youth reporting mild to moderate pain and caregiver reporting no pain. Given

this overall pattern of results (i.e., concurrence of ratings for the majority parent-child dyads, with apparent under-reporting of one caregiver), in order to simplify data analyses only youth self-reports of pain were examined.

There were a total of 595 total diary days recorded during baseline and intervention in this study. Participants reported no pain (i.e., a rating of zero on the visual analog scale) on 70% of the days analyzed; thus, they experienced pain on a total of 30% of days (62% of intervention days). The majority of pain episodes (79%) were rated as mild (1-3 on a 0-10 scale). Twenty-six of the episodes (15%) were rated as moderate pain (4-6), experienced by four youth. Eleven of the episodes (6%), reported by two participants, were rated as severe pain (≤7).

The intensity of pain symptoms decreased for the majority of participants across treatment. Table 3 shows mean pain intensity for days youth were reporting pain. Visual inspection of these scores indicates that Participants 2, 3, 4, 6, and 7 reported improvements between baseline and treatment mean intensity pain ratings. However, response to treatment was variable across participants, with both increases and decreases in pain intensity over the course of treatment.

For Participant 1, of the 82 diary days completed, 70% of the days were pain free (91% during baseline and 55% during intervention). He had considerable variation in pain intensity during the baseline period (see Figure 3). Scheduling treatment proved challenging initially, which resulted in an extended baseline period with stretches of missing data (approximately six weeks). While it appears that from week four to week eight of the baseline phase that pain intensity was on a downward trend, it should be noted that during week seven, Participant 1 was hospitalized for fever and pain crisis, which lasted approximately one week. While daily diaries were not completed for the three weeks prior to treatment, retrospective reports from parent and

child suggest that the participant did not experience any pain in this time frame. As such, it can be presumed that baseline pain intensity (rating of 0) was relatively stable entering into the treatment phase of the study.

After the intervention was implemented there was no change in level of pain intensity (intervention pain intensity rating of 0). While pain intensity showed a decelerating trend during baseline, beginning during week two of treatment an accelerating, steady increase in pain intensity was evident throughout the remainder of the treatment phase. While pain intensity decreased during the final two weeks of the treatment phase, it did not return to the baseline level. This decrease in pain intensity (rating of 2) was not maintained at post-treatment (pain intensity rating of 4). The peak in pain intensity (rating of 5) corresponds with the participant being hospitalized for two distinct pain episodes, with each hospitalization lasting approximately three days. It should be noted that during the last week of treatment, an intense pain crisis was experienced by the participant, but he and his family were able to manage the pain crisis at home, not requiring hospitalization. It appears that Participant 1 demonstrated greater pain intensity at the beginning of baseline and end of treatment, with his lowest pain intensity occurring at the end of the baseline phase and beginning of treatment (first week). Overall, according to overall trend (slope = .06) and mean pain intensity ratings, Participant 1 did not show improvement from baseline to treatment, suggesting that his pain symptoms may not have changed as a function of treatment (i.e., a positive change score of .1; see Table 3).

For Participant 2, of the 93 days completed, 75% of the days were pain free (67% during baseline and 82% during intervention). During the baseline phase, pain intensity remained relatively stable (pain intensity of 1) (see Figure 3). Once the intervention was implemented, a decrease in pain intensity was initially observed. This suggests that an intervention effect was

immediately noted. While pain intensity fluctuated somewhat, periods of stability were noted throughout treatment (i.e., weeks two through five; seven through eight). An overall downward trend of pain intensity was noted across the baseline and treatment phases. In general, this downward trend (slope = -.07) and a rapid latency of change (e.g., the more closely in time that the change occurs after the experimental condition was altered, the clearer the intervention effect) suggest some improvement in pain intensity as a result of the intervention (i.e., a negative change score of .1 was observed from baseline to intervention; see Table 3). This pain intensity rating of 0 was maintained at post-treatment.

For Participant 3, of the 77 days completed, 90% of the days were pain free (71% during baseline and 96% during intervention). During the baseline phase, pain intensity trended upward (initial pain rating of 1 to 2) (see Figure 3). Once the intervention was implemented, a decrease in pain intensity was noted, which suggests that an intervention effect was immediately evident (pain rating of 1). While a peak pain intensity of 1 was noted during week four of treatment, overall, the participant demonstrated a downward trend of pain intensity, as no pain was reported for the remainder of treatment. In sum, mean pain intensity decreased, a rapid latency of change was observed, and a downward trend (slope = -.18) of pain intensity suggests that Participant 3's pain symptoms benefited from intervention (i.e., a negative change score of 1 was observed from baseline to intervention; see Table 3). This pain intensity rating of 0 was maintained at post-treatment.

For Participant 4, of the 56 days completed by the youth, 91% of the days were pain free (81% during baseline and 97% during intervention). During the three week baseline phase, this youth's pain symptoms steadily increased, resulting in a peak pain intensity of 5 (see Figure 3). A rapid decrease in pain intensity was noted once the treatment phase began (to a pain intensity

rating of 0), suggesting a clear intervention effect. Throughout the treatment phase, pain intensity remained relatively stable with no pain being reported for the final three weeks of the intervention. The mean change in pain intensity (i.e., a negative change score of 2 was observed from baseline to intervention; see Table 3) and the rapid change and downward trend (slope = -.53) of pain symptoms once treatment was implemented suggest that this participant's pain symptoms responded favorably to treatment. This pain intensity rating of 0 was maintained at post-treatment.

For Participant 5, of the 56 days completed, 14% of the days were pain free (33% during baseline and 3% during intervention). The low percentage of pain free days for this participant was evident throughout baseline and treatment, as she consistently reported a pain intensity of 1 (see Figure 3). During the baseline phase, no trend of pain symptoms were noted, indicating a relatively stable minimal endorsement of pain (again, baseline pain intensity of 1). When treatment was implemented no change in pain symptoms was evident. During the last week of the treatment phase, the participant reported a slight mean increase in pain intensity (intervention pain intensity rating of 1.6), suggesting an overall upward trend (slope = .06) of pain symptoms from baseline through the intervention. In sum, mean pain intensity ratings increased slightly (i.e., a positive change score of .2 was observed from baseline to intervention; see Table 3), suggesting that the participant did not show improvement from the intervention. However, it should be noted that baseline intensity of pain for this participant was low and thus, there was little room for improvement based on this measure. The mean pain intensity rating of 1 at post-treatment remained unchanged at the 2-month follow-up.

For Participant 6, of the 76 days completed, 26% were pain free (21% during baseline and 38% during intervention). During the baseline phase, a downward trend of pain intensity was

noted (initial mean baseline rating of 3.43 to 2.29) (see Figure 3). When the treatment was implemented, minimal to no change in pain symptoms were initially observed. As the treatment phase progressed, there was some fluctuation of pain symptoms, but the overall downward trend (slope = -.24) suggests that pain decreased throughout treatment (intervention mean pain intensity rating of 1). The participant's mean pain intensity decreased throughout treatment, suggesting some benefit from treatment (i.e., a negative change score of 2.7 was observed from baseline to intervention; see Table 3). However, since the time between the onset of the intervention and decrease in pain symptoms was delayed, it is somewhat less clear that the intervention may have led to the change. The mean pain intensity rating of 1 at post-treatment remained unchanged at the 2-month follow-up.

For Participant 7, of the 77 days completed, 77% of the days were pain free (59% during baseline and 86% during intervention). During the baseline phase, an upward trend of pain intensity was observed (see Figure 3). This steady increase in pain resulted in a peak pain intensity of 8 and an emergency room visit for fever and pain crisis at the end of baseline. When the treatment was implemented, an immediate change in pain intensity was noted (rating of 2 at the beginning of intervention). Throughout the treatment phase, a steady decline in pain symptoms was evident, resulting in a stable trend of no pain from week two of treatment to the end. Overall, Participant 8 demonstrated a decrease in mean pain intensity (i.e., a negative change score of 4 was observed from baseline to intervention; see Table 3), a downward trend (slope = -.35) of pain symptoms, and an immediate response to the treatment intervention, suggesting that he benefited from the treatment intervention. This pain intensity rating of 0 was maintained at post-treatment.

For Participant 8, of the 78 days completed, 95% were pain free (93% during baseline and 95% during intervention). During the two week baseline, an upward trend of pain intensity was noted (e.g., no pain to a minimum rating of 1) (see Figure 3). It should be noted that it is possible that this baseline period was likely not long enough to establish a consistent trend of pain symptoms. With the implementation of treatment, there was no change in pain symptom levels. Fluctuations were noted throughout the treatment phase, with a peak pain intensity of 2 and 1.5 during week four and seven of treatment. As a result, an upward trend (slope = .005) of pain symptoms was noted throughout treatment, and this in conjunction was an increase in mean pain intensity (i.e., a positive change score of 1was observed from baseline to intervention; see Table 3) suggests no benefit to pain symptoms as a result of the intervention. Overall, it appears that her lowest pain intensities were at the beginning of baseline and end of treatment. Her pain intensity rating of 0 was maintained at post-treatment.

Functional Limitations

Functionality During Treatment. Functionality was assessed during intervention using participant responses of daily impairment (i.e., "yes/no" questions) across five dimensions. Indication of impairment (i.e., number of "yes" responses) was summed to create an aggregate score for functionality. For each day, the participant obtained a score ranging from 0 (less dysfunction) to 5 (more dysfunction, activity reduction).

Of the eight participants, three parent-youth pairs had perfect agreement on functioning ratings (see Table 2). The other pairs showed only minor disagreements (parents of Participants 8 and 9 reported less activity than youth self-reports). Since youth and their parents were generally in agreement, only youth self-reports of functioning were plotted.

For all participants, engagement in daily activities according to level of pain was examined. With reports of mild pain (1-3), 84% attended school, 34% participated in activities, 70% were social (e.g., interacted with friends), and 62% completed their chores. With reports of moderate pain (4-6), 56% attended school, 5% participated in activities, 62% were social, and 38% completed their chores. Comparatively, during reports of severe pain (≤7), 0% attended school, 13% participated in activities, and 25% were social and completed their chores.

Table 4 shows mean functioning for the baseline and treatment phases. According to these scores, Participants 2, 3, 4, and 8 reported improvements between baseline and treatment mean functioning ratings. Again, response to treatment was variable across participants and some did not show improvements. Considerable variation was shown from week to week suggesting that participants experienced both increases and decreases in their functioning over the course of treatment. An examination of individual weekly functioning was undertaken to examine changes in means, levels or trends, and latency of change across baseline and treatment.

For Participant 1, functionality during the baseline period appeared to be on a downward trend, despite experiencing a moderate amount of pain in week two (see Figure 4). Again, it should be noted that during week seven of the baseline phase, Participant 1 was hospitalized, which would suggest that his activity was reduced, despite missing diary data. Retrospective reports from parent and child would indicate minimal to no dysfunction after the eighth week of baseline, lending to the assumption that baseline functioning was relatively stable entering into the treatment phase of the study (baseline mean functionality of 0).

After the intervention was implemented there was no change in level of functioning. However, level of dysfunction steadily increased (e.g., trended upward) during the treatment phase, reaching a peak mean dysfunction of 4.71 at week five (which corresponds to the patient

being hospitalized for a pain crisis) before returning to near baseline levels at the end of the treatment phase. Despite a moderate report of pain at post-treatment, Participant1 remained active and seemingly engaged in normal day-to-day activities except for attending school (reported mean functioning of 1). This would suggest an improvement in level of functioning at post-treatment, compared to the preceding treatment weeks. Based upon mean weekly functioning (i.e., a positive change score of 1.5 from baseline to post-treatment; see Table 4) and participant trends (slope = .13), it would appear that this participant did not benefit from treatment.

During the baseline phase for Participant 2 an upward trend was noted, suggesting more activity reduction approaching the treatment phase (see Figure 4). Once the intervention was implemented, less dysfunction was noted almost immediately (mean baseline functioning rating of 1.86 to 1). As a result of this quick response to treatment, a slight upward trend was noted throughout treatment (although, level of functioning remained relatively stable from third week of treatment until the end – mean functioning rating of approximately .5). In general, an overall decrease in mean level of functioning (e.g., less activity reduction) (i.e., a negative change score of .7 from baseline to post-treatment; see Table 4), as well as a overall downward trend (slope = -.04) and rapid latency of change, suggest that the participant received some benefit in level of functioning from the intervention. Reported level of functioning of approximately .5 during the final week of treatment was maintained at post-treatment.

Participant 3 evidenced a slight downward trend during the baseline phase (see Figure 4). Once the treatment phase began, a minor decrease in functional impairment was noted (baseline mean functioning rating of 1.29 to 1), suggesting an immediate intervention effect. However, as treatment progressed a small upward trend (e.g., more activity reduction) was evidenced,

resulting in a peak performance reduction rating of 1.57 during the last week of treatment. While the youth reported mild to no pain, some fluctuation in functioning during the treatment phase was present. Overall, while mean functioning improved slightly (i.e., a negative change score of .2 from baseline to post-treatment; see Table 4), the overall slight downward trend (slope = -.01) suggests minimal benefit with regards to level of functioning for this participant. Upon closer examination of the data, it appears that the participant's level of functioning initially benefited from treatment (e.g., relatively low level of dysfunction for treatment weeks four through eight), but that this improvement tapered as treatment progressed.

Participant 4's level of functioning showed a slight downward trend during the baseline phase (see Figure 4). After the initiation of treatment, an immediate, small change in level (e.g., less activity reduction) was noted (baseline functioning rating of 1.86 to 1.29), suggesting a possible intervention effect. However, as the treatment phase progressed, a steady upward trend was evidenced (level of dysfunction reported at 2), suggesting more dysfunction despite no corresponding reports of pain. For this participant the highest levels of dysfunction were indicated during baseline and at the end of treatment. The participant demonstrated his lowest level of mean weekly dysfunction during the first week of treatment. The overall downward trend (slope = -.04) would suggest some improvement from baseline to post-treatment. Overall, mean functioning evidenced a slight improvement (i.e., a negative change score of .4 from baseline to post-treatment; see Table 4).

During the baseline phase, Participant 5's level of functioning evidenced a downward trend (e.g., less dysfunction) (see Figure 4). When the intervention was implemented, no change in level of functioning was noted. While a slight upward trend appears throughout the treatment phase, the participant consistently lingered around a mean level of functioning of 1 (fluctuations

or inconsistencies were primarily noted in activity participation or spending time with friends).While a linear, upward trend (slope = .02) is noted throughout the study, the participant's lowest levels of dysfunction were evidenced during the last week of baseline and first few weeks of the intervention phase. Overall, mean level of functioning did not benefit from treatment (i.e., a positive change score of .1 from baseline to post-treatment; see Table 4).

During the baseline phase, Participant 6 evidenced a horizontal trend line suggesting a stable level of functioning at approximately one (e.g., minimal dysfunction) despite two weeks of increased pain intensity (see Figure 4). With the implementation of the treatment phase, no change in level was noted. As the treatment progressed, the participant's level of functioning trended upward (e.g., more dysfunction), peaking at week four (mean functioning rating of 2.86. After this peak in reported dysfunction, the participant's level of functioning trended downward, resulting in a return to baseline functioning by the end of treatment (mean functioning rating of 1.29). In sum, Participant 6 evidenced an upward trend (slope = .05) from baseline through treatment, suggesting that her level of functioning did not benefit from the intervention (i.e., a positive change score of .4 from baseline to post-treatment; see Table 4).

Participant 7 indicated more dysfunction (e.g., upward trend) as the baseline phase progressed (see Figure 4), resulting in a peak dysfunction rating of 3.5. As treatment began, the participant's functioning improved (e.g., level decreased) suggesting an immediate benefit from the intervention (initial treatment rating of 2.57). This initial treatment improvement was followed by stable reports of mean functioning, and after a few weeks of treatment, the participant experienced minimal to no activity reductions. However, during the final week of treatment, a steep, upward trend was noted and maintained to post-treatment (mean ratings of 2.86 and 3), which indicated that the participant experienced more dysfunction without changes

in pain (e.g., no pain reported). Given the overall upward trend (slope = .05) and lack of change in mean functioning (i.e., a positive change score of .1 from baseline to post-treatment; see Table 4), this participant's level of functioning did not appear to improve as a result of treatment.

Participant 8 demonstrated a downward trend (e.g., less dysfunction) during the baseline phase (see Figure 4). When treatment began, a slight increase in level was noted (baseline mean functioning rating of .71 to 1), which suggest that any effect from treatment was delayed and may have been confounded by other variables. Following the second week of treatment, the participant experienced a downward trend in level of functioning (e.g., less activity reduction), and reported almost no dysfunction for the remainder of treatment, which continued at post-treatment (maintenance of effect). Changes in functionality did not correspond to reported pain intensity. In general, this participant's mean level of functioning changed (e.g., less dysfunction) (i.e., a negative change score of .7 from baseline to post-treatment; see Table 4) and a downward trend (slope = -.14) was noted throughout, suggesting some possible benefit from treatment.

Functionality Post-Treatment. Table 6 presents the total scores for functionality on the Functional Disability Inventory. Both youth and parent report are presented. Youth and parents only reported concordance at post-treatment and 6-month post-treatment follow-up (correlations were .81 and .97). For Participant 1, adolescent and parent did not report disability at baseline or 4-month post-treatment follow-up (scores = 0). His highest self-reported disability was experienced at post-treatment and 6-month follow-up (scores = 21 and 19 respectively). The only difference between parent and adolescent occurred at the 2-month post-treatment follow-up. While the adolescent reported minimal disability (score = 7), the parent reported moderate symptoms of functional limitations (score = 21).

Participant 2 reported some disability at baseline (score = 13), which reduced at post-treatment (score = 3). A steady increase in disability was noted at 2-month post-treatment (score = 5) and maintained at 4- and 6-month follow-ups (scores = 8). Her parent reported a similar trend of disability, with her highest level occurring at baseline (score = 7), and decreases at post-treatment and 2-month follow-up (scores = 2 and 0 respectively). Then at 4- and 6-month post-treatment follow-up, an increase in disability occurred (scores = 3 and 5 respectively). Participant 3 reported no limitations at baseline, but disability increased at post-treatment (score = 9). A steady decline in functional limitations was noted at 2-month follow-up (score = 2) and decreased to no disability at 4- and 6-month post-treatment. Comparatively, Participant 3's parent reported a moderate amount of disability at baseline (score = 43) which decreased as post-treatment (score = 0) and was maintained throughout the follow-ups.

Participant 4 reported some functional limitations at baseline (score = 11) which decreased at post-treatment and 2-month post-treatment (scores = 2 and 0 respectively). However, at 4- and 6-month follow-up, an increase in disability was noted (scores = 13 and 7 respectively). His caregiver reported minimal to no limitations at all time points except the 2-month post-treatment follow-up (score = 6). For Participant 5, the child and parent reported the highest level of disability at baseline (scores = 8 and 2 respectively), which decreased at post-treatment and throughout the follow-ups (no functional limitations were reported).

For Participant 6, the adolescent and parent reported a steady increase in disability from baseline (scores = 5 and 0) to post-treatment (scores = 8 and 7) to 2-month follow-up (scores = 10 and 3). Participant 7 reported an increase in functional limitations from baseline (score = 2) to post-treatment (score = 17). While a decrease in disability was noted at the 2- and 4-month post-treatment follow-ups (scores = 3 and 0), her highest level of disability was reported at 6-months

post-treatment (score =17). Comparatively, his parent reported a steady increase in disability from baseline (score = 4) to post-treatment (score = 6) to 6-month follow-up (score = 19). For Participant 7, both child and parent reported a decreased in functional limitations from baseline to treatment (scores = 9 and 3 to 6 and 2 respectively).

Health-Related Quality of Life

Post-treatment health-related quality of life scores on the Pediatric Quality of Life Inventory are presented in Table 7. Both youth and parent report are presented. Youth and parents only reported concordance at the 2- and 6-month post-treatment follow-ups (correlations were .82 and .97). For Participant 1, adolescent and parent reported better health-related quality of life at baseline (scores = 75 and 89.13 respectively) which decreased at post-treatment (scores = 54.35 and 57.61 respectively). HRQoL of life improved at 2- and 4-month post-treatment before declining again at the 6-month post-treatment follow-up (scores = 57.61 and 60.87 respectively).

For Participant 2, the child reported an improved HRQoL from baseline (score = 61.96) to post-treatment (score = 89.13). HRQoL fluctuated throughout follow-ups, never reaching as good a score as at post-treatment. Comparatively, her parent reported a decrease in HRQoL from baseline (score = 77.17) to post-treatment (score = 67.39). Parent reported an improvement in HRQoL at 2-month post-treatment (score = 95.65), and while it decreased slightly at the 4- and 6-month follow-up, this score remained higher than initial HRQoL reported. For Participant 3, both child and parent reported better HRQoL from baseline (scores = 67.39 and 39.13 respectively) to post-treatment (scores = 95.65). This improved quality of life was maintained throughout the 6-month post-treatment follow-up. For Participant 4, both adolescent and parent reported an improvement in HRQoL from baseline (scores = 44.57 and 71.74 respectively) to

post-treatment (scores = 98.91 and 95.65 respectively). The adolescent reported a decreased in HRQoL at 2-month post-treatment (score = 61.96); this level of HRQoL was maintained throughout 6-month follow-up. Per parent report, fluctuations in HRQoL were indicated throughout follow-up, with the best HRQoL for the participant being reported at the 4-month follow-up (score = 100).

Participant 5 endorsed an improvement in HRQoL from baseline (score = 59.78) to post-treatment (score = 95.65). This improvement was maintained throughout follow-ups. Her parent reported a decrease in HRQoL from baseline (score = 79.35) to post-treatment (score = 50). An improvement in HRQoL was shown at 2-months post-treatment (score = 93.48) and maintained. For Participant 6, both adolescent and parent reported a slight improvement in HRQoL from baseline (scores = 46.74 and 59.78 respectively) to 2-month post-treatment (scores = 52.17 and 67.39). For Participant 7, both child and parent reported a steady improvement in HRQoL from baseline (scores = 29.35 and 33.70) to post-treatment (scores = 46.74 and 50) to the 6-month post-treatment follow-up (scores = 67.39 and 66.30). While Participant 8 reported a slight decrease in HRQoL from baseline (score = 68.75) to post-treatment (score = 78.26), her parent reported the opposite pattern (i.e., improvement from baseline score of 78.26 to post-treatment score of 86.96).

Psychological Adjustment

Anxiety During Treatment. Anxiety was assessed using the daily diaries. An aggregate score for feelings of state anxiety (i.e., unpleasant emotional state resulting from intensity of pain episode) was created for each day. Participants received a score ranging from 0 (no anxiety) to 20 (high anxiety).

Of the eight parent-youth pairs, only two showed substantial disagreement in mean weekly anxiety ratings. For the cases of disagreement, Participant 5's caregiver reported less daily anxiety than the youth, while there was little to no consistency for Participant 8 during the baseline phases, with parent and youth varying on degrees of anxiety on a daily basis. One parent-youth pairs had perfect agreement on mean anxiety ratings, and the other pairs showed only minor disagreements (see Table 2). Since youth and their parents were generally in agreement, only youth self-reports of functioning were plotted (Figure 5).

Table 5 shows mean functioning for the baseline and treatment phases. According to these scores, only Participants 1 and 3 reported improvements between baseline and treatment mean anxiety ratings. Again, response to treatment was not consistent across participants and some did not show improvements. While there was some variation from week to week, most participants endorsed the same daily levels of anxiety regardless of pain severity (this will be discussed in more detail in the discussion).

An examination of individual weekly functioning was undertaken to examine changes in means, levels or trends, and latency of change across baseline and treatment.

During the baseline phase, Participant 1 endorsed moderate, stable levels of anxiety (mean rating of 8). Again, given the large number of missing data establishing a data trend at baseline was not possible. With the implementation of treatment, mean anxiety level decreased immediately and remained relatively stable and low throughout treatment and at post-treatment (mean anxiety rating of 4) regardless of pain severity. The change in mean weekly anxiety, and overall downward trend (slope = -.35) would suggest the participant's anxiety benefited from treatment (i.e., a negative change score of 1.5 from baseline to post-treatment; see Table 5).

Participant 2 did not endorse any anxiety during the baseline or treatment phases, and as such, determining a treatment effect for anxiety was not possible. Participant 3 demonstrated a downward trend in mean anxiety as the baseline phase advanced. His highest levels of anxiety (mean rating of 6) corresponded to reports of mild pain at week two of the baseline phase. Overall, this participant's level of anxiety did not correspond with severity of pain reports. A downward trend was noted throughout the treatment phase, which resulted in the lowest mean rating of anxiety (score of 1.57) during the fifth week of treatment. The change in mean weekly anxiety, and overall downward trend (slope = -.24) would suggest the participant's anxiety benefited from treatment (i.e., a negative change score of 1.3 from baseline to post-treatment; see Table 5). However, since change in anxiety was somewhat delayed (beginning in week three), attributing this to an intervention effect is not suggested. Other confounds may likely have contributed to this participant's decrease in mean weekly anxiety.

Participant 4 reported moderate anxiety (rating of 8) throughout the baseline and treatment phases. These symptoms did not fluctuate based on pain reports. Overall, the slight upward trend (slope = .02) would suggest that his symptoms of anxiety were not affected by the intervention.

Participant 5 reported minimal anxiety throughout the baseline phase and first few weeks of treatment (anxiety rating of 1). However, an upward trend (e.g., more anxiety) is evidenced during the final weeks of treatment and post-treatment despite no corresponding increases in pain intensity. The overall upward trend (slope = 1.11) throughout the study, and increase in mean weekly anxiety symptoms (i.e., a positive score of 3 from baseline to post-treatment; see Table 5) indicate that this participant's symptoms of anxiety did not benefit from treatment. An unintended negative effect was evident.

Participant 6 reported a moderate level of anxiety (rating of 8) throughout the baseline and treatment phases. These symptoms did not fluctuate based on pain reports. Overall, the slight upward trend (slope = .04) would suggest that her symptoms of anxiety were not affected by the intervention.

Participant 7 reported an upward trend of anxiety throughout the baseline phase. This increase in mean anxiety at baseline (increase in anxiety rating from 11 to 19) corresponds to his severe reports of pain and emergency room visit. When treatment began there was no change in the participant's anxiety level. A horizontal trend line best captures the participant's stable and high anxiety (mean anxiety rating of 20) throughout treatment despite no reports of pain. The lack of change in mean symptoms, overall upward trend (slope = .71), and high level of anxiety indicate that the participant did not benefit from treatment, and unintended effects may have exacerbated his symptoms.

Participant 8 reported a moderate level of anxiety (rating of 12) throughout the baseline and treatment phases. These symptoms did not fluctuate based on pain reports. Overall, the horizontal trend line would suggest that her symptoms of anxiety were not affected by the intervention (slope was not calculated because the participant only endorsed ratings of 12).

Anxiety Post-Treatment. Table 8 presents the raw scores for Total Anxiety Symptoms on the Revised Children's Manifest Anxiety Scale. Participants 4, 6, and 7 endorsed clinically significant anxiety symptoms at various time points (baseline to 6-month post-treatment follow-up). Participant 4 endorsed a clinically significant score at baseline (score = 19) and post-treatment (score = 19). Anxiety symptoms were no longer significant at the 2-, 4-, and 6-month post-treatment follow-up score (scores = 15, 16, and 11 respectively). Participant 6 reported a clinically significant score of 21 at baseline. At post-treatment (score = 18) this score was no

longer significant; however, at the 2-month post-treatment follow-up, she again endorsed a clinically significant score (score = 23). Participant 7 endorsed a clinically significant score at post-treatment (score = 24), but not at baseline (score = 15). While his anxiety symptoms improved at the 2-month follow-up (score = 12), he did not maintain these subclinical symptoms at the 4- or 6-month post-treatment follow-up (scores = 23 and 26 respectively).

Depression Post-Treatment. Table 9 presents the T-scores for Total Depressive Symptoms on the Children's Depression Inventory, as well as at-risk or clinically significant subscales endorsed by participants. Specifically, Participants 4, 5, 6, and 7 demonstrated at-risk and clinically significant subscales at various time points (baseline to 6-month post-treatment follow-up). Participant 4 endorsed a clinically significant score for Negative Self-Esteem at baseline (T=70). At post-treatment (T=45) and the 2-month post-treatment follow-up (T=40) his self-esteem improved, as his score was subclinical. While his negative self-esteem score, at the 4-month follow-up was again in the at-risk range (T=60), it returned to the subclinical level at the 6-month post-treatment follow-up (T=45). Participant 5 reported an at-risk concern for Anhedonia at baseline (T=65). By post-treatment this score was subclinical and remained as such throughout the 6-month post-treatment follow-up (T=38). At baseline, Participant 6 endorsed an at-risk concern for Total Score (T=62) and Ineffectiveness (T=66). Ineffectiveness was subclinical at post-treatment (T=59) and such an effect was maintained at 2-month post-treatment follow-up (T=59). While her total depressive symptoms improved at post-treatment (T=50), it again reached the at-risk range at the 2-month post-treatment follow-up (T=62). At baseline, Participant 7 endorsed an at-risk concern for Anhedonia (T=64). Anhedonia remained in the at-risk range at post-treatment (T=64). This score was subclinical at the 2- and 4-month

post-treatment follow-ups (T=52 and 48 respectively), but was again in the at-risk range at 6-months post-treatment (T=60).

Coping Strategies Post-Treatment. Table 10 presents the composite factor scores for the Coping Strategies Questionnaire. Participant 1 demonstrated an improvement in Coping Attempts from baseline (score = 51) to post-treatment (score = 64). However, he did not appear to maintain these active coping strategies at the 2- or 4-month follow-ups (scores = 49 and 28), but reported greater use at the 6-month post-treatment follow-up comparable to his post-treatment score. He reported a decrease in Negative Thinking and Passive Adherence from baseline (scores = 58 and 88) to post-treatment (scores = 41 and 74) to 4-month post-treatment follow-up (scores = 33 and 62). Participant 2 reported an improvement in Coping Attempts from baseline (score = 94) to post-treatment (score = 112) to 6-month post-treatment follow-up (score 124). While a decrease in Negative Thinking from baseline (score = 33) to post-treatment (score = 7) was noted, this improvement was maintained (no reported increases or decreases) throughout follow-ups. No change in Passive Adherence was reported.

Participant 3 demonstrated an improvement in Coping Attempts from baseline (score = 60) to post-treatment (score = 89) to 2-month follow-up (score = 95). This improvement in Coping Attempts was not maintained. He indicated a decrease in Negative Thinking from baseline (score = 36) to post-treatment (score = 26). While an increase was noted at the 2- and 4-month follow-up, his lowest score was reported at the 6-month post-treatment follow-up (score = 8). Passive Adherence steadily increased from baseline (score = 57) to post-treatment (score = 81) to 2-month follow-up (score = 92). Participant 4 demonstrated an increase in active Coping Attempts and Passive Adherence from baseline (score = 39 and 63) to post-treatment (score = 78 and 76); at the 4-month post-treatment follow-up active Coping Attempts continued to increase

(score = 127). In conjunction with these improvement, he endorsed an increase in the Negative Thinking as well from baseline (scores = 48 and) to post-treatment (scores = 73).

While Participant 5 endorsed a decrease in active Coping Attempts from baseline (score = 74) to post-treatment (score = 54), her Negative Thinking also decreased from baseline (score = 92) to post-treatment (score = 15). However, her Negative Thinking steadily increased throughout the 2-, 4-, and 6-month post-treatment follow-ups, but never reached the high baseline level of this negative coping strategy. While Passive Adherence decreased from baseline (score = 87) to post-treatment (score = 78), it steadily increased throughout the follow-ups. Participant 6 demonstrated a decrease in active Coping Attempts from baseline (score = 84) to post-treatment (score = 68) to 2-month follow-up (score = 61). A decreased was also noted for Passive Adherence coping strategies from baseline (score = 85) to post-treatment (score = 68). On a positive note, her Negative Thinking also steadily decreased from baseline (scores = 72) to post-treatment (scores = 44).

Participant 7 endorsed an increase in active Coping Attempts and Passive Adherence from baseline (score = 76 and 94) to post-treatment (score = 158 and 132). His active coping strategies were maintained at 2-months post-treatment before decreasing at the 4-month follow-up (score = 41). Comparatively, Negative Thinking increased from baseline (scores = 62) to post-treatment (scores = 100). Participant 8 indicated an increase in Coping Attempts, Negative Thinking, and Passive Adherence from baseline (score = 48, 35, and 78) to post-treatment (score = 89, 76, and 103).

Spiritual Coping. Table 11 presents the total scores for Religious and Existential Coping Frequency and Efficacy from the Child Spirituality Coping Survey. Participant 1 endorsed a decrease in religious coping strategies, frequency and efficacy, from baseline (scores = 29 and

28) to post-treatment (scores = 11 and 12). However, the frequency and efficacy of these religious strategies increased at 2- and 4-month follow-up before decreasing at 6-month post-treatment (scores = 6). Existential coping strategies, frequency and efficacy, steadily increased from baseline (scores = 10) to post-treatment (scores = 12) to 4-month follow-up (scores = 24 and 18 respectively). However, at the 6-month follow-up, he endorsed the lowest frequency and efficacy scores (scores = 8 and 4). While Participant 2 indicated an increase in religious coping strategies frequency from baseline (score = 18) to post-treatment (score = 24), the believed efficacy of these strategies decreased (baseline score of 21 to post-treatment score of 18). The frequency and efficacy of these religious coping strategies further decreased throughout the various follow-ups. Similar decreases in existential coping strategies, frequency and efficacy, were shown from baseline (scores = 36 and 32) to post-treatment (scores = 32 and 30) to 4-month follow-up (scores = 17 and 16). However, at the 6-month follow-up this participant again endorsed similar use and perceived effectiveness of these existential coping strategies at baseline.

Participant 3 did not endorse the use of any religious or existential coping strategies at baseline. As such the use and efficacy of these strategies, both religious and existential, increased at post-treatment (scores in mid-to-upper 20s). He continued to endorse similar religious and existential coping strategies throughout the follow-ups. While Participant 4 indicated a slight decrease in religious coping frequency from baseline (score = 28) to post-treatment (score = 26), the frequency increased at the 2-month follow-up (score = 34) and maintained throughout the remainder of the follow-ups. Religious coping efficacy mirrored these same trends. Comparatively, he endorsed an increase in existential coping, frequency and efficacy from baseline (scores = 29 and 19) to post-treatment (scores = 35). The frequency of these strategies

peaked at the 2-month follow-up (score = 52) and steadily decreased throughout the remainder of the follow-ups. Participant 5 did not report a change in religious coping strategies; however, a steady increase in existential coping strategies, both frequency and efficacy, was endorsed from baseline (scores = 33 and 34) to post-treatment (scores = 38) to 4-month post-treatment follow-up (scores = 52).

Participant 6 did not endorse any changes in religious or existential coping strategies from baseline to 2-month follow-up. Participant 7 endorsed an increase in the efficacy of religious coping strategies from baseline (score = 25) to post-treatment (score = 36). This change was maintained throughout the 2-, 4-, and 6-month post-treatment follow-ups. He also endorsed an increase in existential coping strategies, both frequency and efficacy from baseline (scores = 42 and 28) to post-treatment (scores = 48). These changes were not maintained at follow-up, but did approach post-treatment scores at 6-month follow-up. Participant 8 did not endorse any changes in religious or existential coping strategies from baseline to post-treatment.

Internalizing/Externalizing Symptoms Post-Treatment. Table 12 presents the T-scores for Internalizing and Externalizing symptoms and Total Behavior Problems on the Child Behavior Checklist. Specifically, the caregivers of Participants 1, 2, 3, 6, and 7 endorsed at-risk and clinically significant scores at various time points (baseline to 6-month post-treatment follow-up). The parent of Participant 1 did not endorse any clinically significant or at-risk concerns at baseline or post-treatment. However, at the 2-month follow-up, an at-risk concern for Internalizing symptoms was reported (T=61). This was no longer an at-risk concern at the 4- or 6-month post-treatment follow-up. The parent of Participant 2 endorsed a clinically significant concern for internalizing symptoms at baseline (T=65) and post-treatment (T=68). At the 2-, 4-, and 6-month follow-up, his internalizing symptoms were reported to be subclinical. An at-risk

concern was endorsed at baseline (T=63) for total behavior problems, and at post-treatment this score became clinically significant (T=65). For the 2-, 4-, and 6-month follow-ups Total Behavior Problems were reportedly subclinical. The parent of Participant 3 indicated an at-risk concern for internalizing symptoms at baseline (T=61). By post-treatment this score was subclinical and remained as such throughout the 2-month post-treatment follow-up (T=48). The parent of Participant 6 endorsed an at-risk concern for internalizing symptoms at baseline (T=60) and post-treatment (T=60). By the 2-month follow-up this score was subclinical (T=50). The parent of Participant 7 endorsed clinically significant concerns for internalizing symptoms at baseline (T=70) and post-treatment (T=67). This concern for internalizing symptoms was subclinical by the 2-month follow-up and remained as such throughout the 6-month post-treatment follow-up. In addition, a clinically significant concern for total behavior problems was also endorsed at baseline (T=64). By post-treatment, this score was no longer significant (T=59) and remained as this throughout post-treatment follow-ups.

Parenting Behaviors. Table 13 presents the factor scores for the Adult Responses to Children's Symptoms measure. Participant 1's caregiver endorsed decreases in Protecting and Distracting and Monitoring mean behaviors from baseline (scores = 2 and 3.13) to post-treatment (scores = 2.40 and 2.88) to 4-month post-treatment follow-up (scores = .80 and 1.5). This positive change in parenting behavior was not maintained at the 6-month follow-up. While Participant 2's parents did not demonstrate a change in Protecting behaviors from baseline to post-treatment (score = 2), a decrease was noted at 2-month follow-up (score = 1.2) and maintained for the additional post-treatment assessments. Distracting and Monitoring parenting behaviors decreased from baseline to post-treatment to 2-month follow-up. This improvement was not maintained at the 4- or 6-month follow-ups.

Participant 3's caregiver reported a decrease in Protect and Distract and Monitor behaviors from baseline (scores = 3) to post-treatment (scores = 1.93). Only the decrease in protecting behavior was maintained at the 2-month follow-up. The parent of Participant's 4 and 5 demonstrated an increase (or maintained these problematic parenting behaviors) for Protecting and Monitoring behaviors throughout the intervention and follow-ups. Participant 6's caregiver only demonstrated a decrease in Monitoring behavior from baseline (score = 2.13) to post-treatment (score = 1.38) (which was also maintained at the 2-month post-treatment follow-up). The parent of Participant 7 reported a decrease in Protecting and Monitoring behaviors from baseline (scores = 3.33 and 3.75) to post-treatment (scores = 2.40 and 3.38). The engagement in Monitoring behaviors further decreased at 2-months (score = 2.63) before returning to baseline levels at the 4- and 6-month follow-ups. Participant 8's parent endorsed a decrease in Protecting and Monitoring behaviors from baseline (score = 3.8 and 4) to post-treatment (scores = 2.87 to 3.13). It should be noted that for all participant's no changes in Minimizing behaviors was reported, likely because baseline levels were already quite low for this problematic parenting style.

Parenting Stress. Table 14 presents the total frequency and difficulty scores for the Pediatric Inventory for Parents. Higher scores indicate greater frequency of problems in the areas of communication, medical care, emotional disturbance, and role function, as well as more difficulty in these areas. Participant 1's parent endorsed an increase in the frequency and difficulty of problems from baseline to post-treatment. Fluctuations were noted throughout post-treatments. The caregiver of Participant 2 also reported an increase in the frequency and difficulty of problems associated with their child's SCD from baseline to post-treatment, but decreases in the 2-, 4-, and 6-month follow-ups were noted. While the parent of Participant 3

endorsed a decrease in the frequency and difficulty of problems from baseline to post-treatment, and elevation was noted in difficulties at the 2-month follow-up.

While the caregiver of Participant's 4 and 5 endorsed an increase in the frequency of problems between baseline and post-treatment, the difficulties associated with these problems decreased in the same time frame. Participant 6 reported an increase in the frequency and difficulties associated with their child's SCD from baseline to post-treatment. The caregiver of Participant 7 endorsed a decrease in the frequency and difficulties associated with such problems from baseline to post-treatment to 6-month post-treatment follow-up. Participant 8 reported an increase in the frequency and difficulties associated with their child's SCD from baseline to post-treatment.

Health Care Utilization. Across participants, hospitalizations during treatment appeared to be generally consistent with those during intervention (Table 15). Participants who required hospitalization for pain episodes prior to intervention also required this during intervention and/ post-treatment. However, anecdotally, several participants noted to the clinician feeling that they had been able to manage a pain crisis during treatment and avoid hospitalization.

Exploratory Statistical Analysis of Treatment Effects

Although statistical analyses of treatment change during and following intervention were limited given the small sample size, general linear modeling was conducted to explore the long-term effects of treatment. A repeated-measures general linear model analysis was undertaken. Separate analyses were run for all outcomes (i.e., pain, functionality, HRQoL, anxiety, depression, internalizing/externalizing symptoms, coping strategies, and parenting behaviors and stress) that were measured on five occasions (e.g., baseline, post-treatment, and 2-, 4-, and 6-

month post-treatment follow-ups). Given the small sample size, no between-subject variables were controlled for. The magnitude of changes from baseline to post-treatment, and baseline to 2-, 4-, and 6-months post-treatment were examined by calculating effect sizes (Cohen's *d*), which reflect the difference between means divided by standard deviation while accounting for sample size[1]. Given within-subjects analyses, the dependence among means was corrected for by including mean correlations.

No significant main effect for time was found for pain symptoms (as measured on the PPQ) or functionality (as measured on the FDI). Per child report, overall mean functionality demonstrated fluctuations throughout baseline and treatment, with the highest level of disability reported at the 6-month post-treatment follow-up (*M*=10.20). Parent's reported a mean decrease in disability from baseline (M=7.63, SD=14.47) to post-treatment (M=5, SD=7.35, *d* =.211) to 4-month follow-up (M=1.83, SD=3.25, *d*=.517); however, similarly to youth reports, the highest level of disability was again noted at the 6-month follow-up (M=9).

A significant main effect of time was found for youth-reported HRQoL, $F(4, 20) = 4.6$, p=.01, *d*=-1.021. No significant main effect of time was found for the parent's report of youth HRQoL (as measured on the PedsQL). Based on child and parent-report, all participant's HRQoL improved from baseline (M=56.69, SD=15.24 and 66.03, SD=20.09) to post-treatment (M=74.46, SD=22.58, *d*=.25 and 69.67, SD=19.80, *d*=-.145) to 4-month post-treatment (M=86.59, SD=14.95, *d*=-2.716 and 86.69, SD=14.04, *d*=-1.170). A slight decrease was noted in HRQoL at the 6-month post-treatment follow-up, as compared to the 4-month follow-up; however, 6-month scores were still better than baseline reports.

[1] Guidelines for interpreting effect sizes is as follows: $d = 0.20$ indicates a small (but not trivial) effect, $d = 0.50$ indicates a medium effect, and $d = 0.80$ indicates a large effect.

No significant main effect for time was found for anxiety symptoms (as measured on the RCMAS). Although not significant, overall mean anxiety scores decreased throughout treatment for all participants. Baseline anxiety symptoms (M=12.27, SD=5.53) decreased at post-treatment (M=10.38, SD=8.98, *d*=.187) and further reduced at the 6-month post-treatment follow-up (M=8.33, SD=9.24, *d*=.465).

A significant main effect of time was found for depressive symptomatology, $F(4, 20) = 4.5$, $p=.01$, $d=1.087$. For all participants, overall mean scores (as measured on the CDI) decreased throughout treatment and follow-up. Specifically, baseline depressive symptoms (M=48.17, SD=8.64) decreased at post-treatment (M=43.33, SD=8.14, *d*=.572) and further reduced at the 6-month post-treatment follow-up (M=40.5, SD=5.99, *d*=1.087).

According to parent-reports (as measured by the CBCL), a significant main effect of time was found for Internalizing, $F(4, 16) = 3.4$, $p=.03$, $d=1.802$, Externalizing, $F(4, 16) = 7.2$, $p=.00$, $d=1.851$, and Total Behavior Problems, $F(4, 16) = 7.7$, $p=.00$, $d=8.268$. The overall mean scores for all participants decreased throughout treatment and follow-ups. Baseline internalizing symptoms (M=56.38, SD=11.39) decreased at post-treatment (M=52, SD=13.27, *d*=.494) and further reduced at the 6-month post-treatment follow-up (M=42.2, SD=10.43, *d*=1.625). Baseline externalizing symptoms (M=51, SD=6.85) decreased at post-treatment (M=44.88, SD=8.81, *d*=1.408) and further reduced at the 6-month post-treatment follow-up (M=39.2, SD=7.43, *d*=1.911). Lastly, baseline total behavior problems (M=53.88, SD=8.18) decreased at post-treatment (M=48.63, SD=10.35, *d*=.864) and further reduced at the 6-month post-treatment follow-up (M=37.8, SD=10.23, *d*=7.382).

No significant main effect for time was found for general or spiritual coping strategies. On the CSQ, Coping Attempt mean scores increased from baseline (M=65.75, SD=19.22) to

post-treatment (M=89, SD=33.17, *d*=-0.751) to 6-month post-treatment follow-up (M=91.60, SD=29.33, *d*=-.914) for all participants. Decreases in the positive coping strategy were noted at the 2- and 4-month follow-ups. Negative Thinking mean scores decreased from baseline (M=54.50, SD=20.73) to post-treatment (M=47.75, SD=32.58, *d*=.186). This decrease in negative thinking was maintained throughout follow-ups (e.g., not further improved upon). Passive Adherence (e.g., adherence to physicians' recommendations including resting and fluid intake) mean scores increased from baseline (M=80.13, SD=13.36) to post-treatment (M=88, SD=20.93, *d*=-.406) to 6-month post-treatment (M=92, SD=18.23, *d*=-1.386); however, a decrease in this strategy was noted at the 2-month follow-up (M=77.50, SD=40.42, *d*=.081). It should be noted that affective coping strategies (i.e., Negative Thinking scale) is considered psychologically inappropriate, while Passive Adherence strategies consist of coping strategies that are considered useful for medical purposes. Active Coping Attempts are psychologically the most positive strategy a youth can utilize. On the CSCS, religious coping mean frequency and efficacy scores did not demonstrate change throughout treatment or the post-treatment follow-ups. Participants did report a minimal increase in existential coping mean frequency and efficacy scores from baseline (M=25.75, SD=13.92 and 22.50, SD=12.02) to post-treatment (M=31, SD=10.43, *d*=-.552 and 30.5, SD=11.24, *d*=-.781), which were also maintained throughout the follow-ups. Across all time points, mean item responses for both religious and existential coping strategies fell in the middle of the scale between "you do this sometimes" and "you do this a lot."

No significant main effect for time was found for parenting behaviors or stress. Mean parenting behaviors, across the three scales (Protect, Minimize, Distract and Monitor), did not change from baseline to post-treatment, suggesting minimal benefit from the intervention. The mean frequency and difficulty of communication, medical care, role function, and emotional

disturbance increased for parents from baseline to post-treatment; however, a steady decline in the difficulties associated with their child's SCD decreased throughout the 2-, 4-, and 6-month post-treatment follow-ups.

Intention-to-Treat Analyses

For controlled trials, intention-to-treat analysis – the inclusion of all participants in the analysis according to group determined at randomization – has become the standard. As part of this analysis, methods are used to estimate the missing data for patients who have dropped out. One such method is the "last observation carried forward (LOCF)." This technique replaces a participant's missing values after dropout with the last available measurement. This assumes that the participant's responses (e.g., outcome measures) would have been stable from the point of dropout to trial completion, rather than declining or improving further (Unnebrink & Windeler, 2001). It also assumes that the missing values are "missing completely at random" (i.e., that the probability of dropout is not related to variables such as disease severity, symptoms, etc.) (Gadbury, Coffey, & Allison, 2003). The advantages to this approach are that (a) it minimizes the number of subjects who are eliminated from the analysis, and (b) it allows the analysis to examine trends over time, rather than focusing simply on the endpoint (Mallinckrodt et al., 2003).

Using the LOCF method for Participants 6 (dropout at 4- and 6-month post-treatment follow-up) and 8 (drop out at the 2-, 4-, and 6-month post-treatment follow-up) a repeated-measures general linear model analysis was completed again as described above, with separate analyses for all outcomes (i.e., pain, functionality, HRQoL, anxiety, depression, internalizing/externalizing symptoms, coping strategies, and parenting behaviors and stress) that

were measured on five occasions (e.g., baseline, post-treatment, and 2-, 4-, and 6-month post-treatment follow-ups).

In general, the intention-to treat LOCF analyses (n=8) were quite consistent with the main analyses described above (utilizing listwise deletion; n = 6). Consistent with the exploratory findings, the intention-to-treat analyses also did not demonstrate a significant main effect for time for pain symptoms (as measured on the PPQ), functionality (as measured on the FDI), anxiety symptoms (as measured on the RCMAS), or general and spiritual coping strategies. A significant main effect of time was also maintained in the intent-to-treat analysis for youth-reported HRQoL, $F(4, 28) = 4.0$, $p=.01$, depressive symptomatology, $F(4, 23) = 3.5$, $p=.05$ (as measured on the CDI), as well as for parent-reports (as measured by the CBCL) of Internalizing, $F(4, 28) = 5.5$, $p=.01$, Externalizing, $F(4, 28) = 11.5$, $p=.00$, and Total Behavior Problems, $F(4, 28) = 8.1$, $p=.00$. However, two differences were observed in the intention-to-treat analyses. First, a significant main effect of time was found for parent's report of youth HRQoL $F(4, 28) = 2.9$, $p=.05$ (as measured on the PedsQL) with improvements observed from baseline to the 6-month post-treatment follow-up. Secondly, a significant main effect of time was also found for parenting behaviors related to monitoring SCD (Distract and Monitor) $F(4, 28) = 3.0$, $p=.05$, with parents engaging in less question asking and fewer checking behaviors from baseline to the 6-month post-treatment follow-up.

Discussion

The primary objective of this study was to evaluate the feasibility of a modified cognitive-behavioral family intervention (CBFT) designed to simultaneously target pain symptoms, functionality, HRQoL, and psychosocial symptoms associated with pediatric SCD.

The results of this pilot study suggest the benefit of this culturally sensitive and family based CBT approach for youth with SCD. Importantly, as a group, participants evidenced statistically significant decreases in psychosocial symptoms (i.e., depression, internalizing and externalizing behaviors) and increases in HRQoL. These improvements in psychosocial symptoms and health outcomes were generally maintained at the 6-month post-treatment follow-up. Furthermore, five of the eight youth also demonstrated decreases in pain symptoms from baseline to post-treatment. With regards to coping strategies, increases in active coping attempts and adherence to medical recommendations were observed from baseline to post-treatment; however, fluctuations in each participant's application of these strategies were noted at follow-up. In addition, negative thinking decreased from baseline to post-treatment and this positive change in coping was maintained through the 4-month follow-up. Overall, these results provide some support for the study hypotheses and suggest that this intervention has potential for broad ranging benefits for pediatric patients with SCD.

Indications of Intervention Gains

Importantly, the CBT intervention was associated with a number of positive outcomes for pediatric patients with SCD in a variety of domains, including reductions in pain symptoms, improvements in functionality, psychological adjustment outcomes, coping strategies, and HRQoL. These improvements were observed on visual inspection of the data (i.e., the trend lines of the data), by statistical analyses (when appropriate), and by examination of effect sizes. However, it is important to interpret these results cautiously given the limitations inherent to these approaches (discussed below). Furthermore, given that limited stability (i.e., defined as a stable, upward, or downward trend evidenced by 2 to 3 consistent weeks of identical ratings) was

observed during baseline assessment, it is also necessary to cautiously interpret the changes observed. A lack of baseline stability prior to intervention limits the ability to draw conclusions that change observed is the result of intervention.

First, reductions in the intensity of pain symptoms during the intervention were noted for five of the eight participants (Participants 2, 3, 4, 6, and 7). Based on suggested benchmarks for evaluating the magnitude of changes in pain intensity (Dworkin et al., 2008), two of the participants "improved" (i.e., decrease in pain intensity less than 2 points or <30%), two were "much improved" (i.e., decrease in pain intensity of 2 points or 30%), and one participant was "very much improved" (i.e., decrease of ≥4 points or ≥50%) in pain symptom reductions. However, it should be noted that half of the participants were receiving Hydroxyurea therapy, which has previously been shown to reduce the rate of acute pain episodes for a one third of the patients who receive it. Therefore, changes in pain are to be cautiously interpreted given this confounding variable.

Second, improvements in functional limitations were observed for four of the eight participants (Participants 2, 3, 4, and 8), although these changes were minimal. Participant report of medication use and activity reduction on pain days was inconsistent. For example, Participants 1 and 7 took medication (e.g., ibuprofen, Tylenol with codeine) on 88% and 61% of days they experienced pain. In contrast, Participants 2 and 6 only took medication on 20% and 7% of days they experienced pain. Furthermore, as pain intensity increased functionality decreased (i.e., decreased school attendance, participation in activity and completion of chores). Previous studies have shown a somewhat consistent pattern in the way SCD pain is related to pain response, with higher levels of pain related to greater medication use, increased health care use, and greater activity reduction (Gil et al., 2000). While the adult literature demonstrates continued work or

school attendance while in pain, the child literature demonstrates less consistent findings. Specifically, although some children are more likely to miss school on pain days, other children never miss school despite pain episodes (Gil et al., 2000). Thus, given the results of the present study, which are in line with the existing literature, it is not possible to draw conclusions about the relationship between pain and functionality.

Third, evidence of reduction in anxiety symptoms was evident from the daily diary reports for two participants (1 and 3) and at post-treatment. Overall, report of daily anxiety for all participants appeared to be independent of pain intensity. It was expected that daily reports would capture state anxiety (i.e., a temporary, unpleasant emotional reaction when faced with a frightening or challenging stressor such as pain). However, these daily reports seem to more accurately reflect trait anxiety (i.e., perpetual feelings of persistently expectedly bad circumstances to transpire regardless of the situation). For example, throughout the intervention phase Participant 8 consistently reported the highest level of anxiety despite no report of pain. In comparison to the daily dairy measure, participant anxiety ratings at post-treatment on the RCMAS generally demonstrated decreases in anxiety (post-treatment and at 2-month follow-up), although these changes were not statistically significant. It should be noted that the daily dairies queried anxiety as a function of pain (i.e., "I worry when I am in pain") whereas the RCMAS may have provided a more general measure of anxiety related symptoms (i.e., "I get nervous when things do not go the right way for me"). It may be that the daily report of anxiety was not sensitive as a measure of changes in anxiety related to pain experience. The act of repeatedly (daily) responding to the same statements may not have captured subtle changes in anxiety-related perception of pain.

Fourth, results of the present study indicate that the intervention significantly decreased depressive symptoms (as reported by the child), as well as total behavior problems (as reported by parents) from baseline to post-treatment, and these results were maintained throughout the 6-month follow-up. While it is important to note these improved symptoms, most participants did not have scores in the clinically significant range on the CDI. However, for the two participants who endorsed suicidal ideation at baseline, these participants did not endorse these feelings at any of the post-treatments. Studies which have examined psychosocial adaptation of children and adolescents with SCD have reported variable levels of functioning, although more internalizing symptoms tend to be observed (i.e., depression, somatic complaints, anxiety) in comparison to control groups and youth with other chronic illness (Barakat, Schwartz, Simon, & Radcliffe, 2007). In contrast, although some children and adolescents with SCD experience behavior problems, these are generally not reported at a higher rate than for peers (Thompson et al., 1993).

Fifth, the results of the present study suggest that, while not statistically significant, all participants appear to demonstrate an increase in active coping strategies from baseline to post-treatment and that this was maintained through the 2-month follow-up. That is, on visual inspection the data appear to trend toward an increase. Participants also reported an increase in passive adherence from baseline to post-treatment. Previous research has shown that passive adherence to medical recommendations (such as taking fluids and resting) may be an effective means of coping but only when other more adaptive strategies are also used (Gil et al., 1989). When only physiological strategies are used, the risk is for becoming inactive and dependent upon health care services for pain management. This pattern appeared to be evident at the 4-month follow-up, as use of passive coping strategies increased, while active coping attempts decreased. In comparison, negative thinking decreased (considered a positive adjustment in

coping strategy utilization) from baseline to post-treatment and was maintained at the 4-month follow-up. Previous research has shown that lower levels of negative thinking are associated with better adjustment to SCD (e.g., fewer health care contacts, and less psychological distress) (Gil et al., 1989, 2001).

These generally positive changes in daily use of coping skills may be associated with more successful home management of pain in children with SCD. However, it is important to note that although gaining control over pain symptoms (i.e., altering negative perception regarding ability to manage pain and the psychological consequences of pain) was a target of the intervention, this was not explicitly measured. As such, no conclusions can be drawn about perceived self-efficacy, and whether thoughts equate into behavior change or action. As such, although no conclusions can be drawn about the relationship between perceived self-efficacy and behavioral changes it is likely that increases in active coping attempts would be associated with improved pain management.

Clearly, many factors contribute to how a child copes with chronic illness. Given the salience of faith in the African-American community, it was anticipated that youth would report using religion and prayer to manage pain and other difficulties associated with SCD. All participants reported a consistently high to moderate amount of religious coping strategies, such as turning to prayer or using their spiritual beliefs for symptom management. No changes were noted throughout the intervention, indicating that religious coping strategies were utilized consistently, in addition to general coping strategies. It should be noted that changes in religious and spiritual coping strategies were not predicted. Rather, these coping strategies were of interest in the present study, given the culturally sensitive approach, but were not specifically targeted during the intervention. At baseline, all but one participant endorsed religious or

existential coping strategies (and all participants endorsed religious/existential strategies at post-treatment). This is consistent with other research which has found that religious and spiritual beliefs allow individuals to gain a sense of control over their illness, construct meaning out of their experiences, and achieve a feeling of comfort and closeness to the Divine (Cotton, Grossosehme, & McGrady, 2011). In the current study, it appears that pediatric patients used a variety of coping means, including religious and more general strategies, to adjust to pain and various stress associated with their illness. However, in contrast to use of general coping, use of religious strategies was consistent and frequent, which is an important consideration from an intervention standpoint.

Sixth, the present intervention was effective in improving HRQoL. Youth reported statistically significant improvements in their activity level, peer relationships, school functioning, and emotional adjustment. This stands in contrast to the minimal improvement on measures of functionality (as described above) and suggests the possibility of some gains in this domain. This suggests that although participants may not have experienced changes in their perception of the impact of their pain on more specific indices of their functionality, they nonetheless experienced improvements in ways that were captured by more broad indices including their relationships and emotional adjustment.

Seventh, although family functioning may be a powerful determinant of overall quality of life for youth with chronic conditions, changes in this domain were not observed as a function of the intervention. No significant changes were noted in parental responses such as minimizing (e.g., shifting focus away from pain behaviors, expressing negative or punishing reactions to child's pain) or encouragement/reinforcement (e.g., frequent attending to pain symptoms, granting permission to avoid regular activities) (Petersen & Palermo, 2004). Parental stress (as

measured by the PIP) also did not change as a function of the intervention. More specifically, the frequency of stressful events was variable but increased slightly throughout the intervention and follow-up. Although the goal of the present study was to incorporate family-based components into the intervention, this was accomplished by placing the caregiver in a role supportive to the child's treatment. In other words, although caregiver concerns and challenges were broadly addressed as they arose during the course of treatment, caregivers did not receive an intervention which specifically targeted their own needs related to their child's SCD. Therefore, it may be relevant to consider how specific parent variables could and should be targeted within the context of this intervention. For example, the present treatment focused on providing strategies for youth, and encouraging parents to support and encourage their children in this process. While this may have been a successful fit for some parents, others may have benefited from a treatment which more actively addressed their own challenges and concerns related to their child's SCD and related functioning.

Contributions and Clinical Considerations

The primary strength of the current study was to extend a CBT pain intervention for RAP to the pediatric SCD population, while implementing culturally sensitive components expected to enhance retention and the effectiveness with an African American population. Following a review by Schwartz et al. (2007) which considered culturally sensitive components when working with African American families, the following strategies were incorporated into the present study: (a) emphasizing the role of the family to develop realistic and feasible coping strategies, (b) emphasizing empowerment or gaining a sense of control for managing pain, (c) including content that was culturally sensitive (e.g., self-statements, supportive material, and

flexible language chosen by youth), (d) awareness of stigma of mental health problems (e.g., normalizing not pathologizing problems, support and reinforce openness, disclosure, and help-seeking behaviors), and (e) allowing flexibility in scheduling. Given that all eight participants who began the treatment were able to complete the 5-session intervention (and six of these completed all follow-up assessments), it appears likely that these culturally sensitive modifications contributed to this success rate.

As a primary consideration of the present study, the 5-seession cognitive behavioral family-based treatment (CBFT) was demonstrated to be feasible as an intervention for the SCD population. This was evidenced by the participation rate of families in attending all sessions over the course of a 3-month period, as well as the minimal dropout rate. While several families were unable to participate in the study due to financial and transportation difficulties, such a limitation is consistently noted in SCD studies unless community or home-based interventions are provided. This study also appeared to be well received by participants, according to the program evaluations completed at the time 2 post-intervention. All youth and their parents reported that they would "recommend" or "strongly recommend" the intervention to a friend. Seven of the eight youth reported feeling "satisfied" or "very satisfied" with their progress, while all parents endorsed these statements. With regards to techniques of coping, seven of the eight participants reported "several" and "many" new techniques. Furthermore, all participants reported "somewhat more" or "much more" confidence in their ability to cope with pain. These highly positive ratings are consistent with other research which has reported higher satisfaction with culturally adapted interventions (e.g., Schwartz et al., 2007; Martinez & Eddy, 2005).

Another strength is the use of a multiple baseline design to examine changes in pain symptoms, functioning, and anxiety in individual participants. Evaluation on these dimensions

occurred on a daily or weekly basis, yielding a detailed characterization of intervention effects, in contrast to group designs which may not allow such a fine-grained approach (Kazdin, 2010). An additional strength was the use of prospective diaries rather than retrospective interviews, which increases the validity of pain reports (Gil et al., 1997). Unlike retrospective methods, prospective diaries do not require a summary or average of behaviors and can elicit more accurate descriptions (Palermo, Valenzuela, & Stork, 2003).

Although results of the present study were based on youth report, the present study prompts some consideration of the relationship between parent and child reports. Current recommendations suggest a multi-informant approach using self and caregiver report of pain for daily diaries, as research suggests that youth reports of pain are generally, but not consistently, concordant with those provided by caregivers (Barakat, Simon, Schwartz, & Radcliffe, 2008). This was evident in the present study, as the majority of youth-parent daily-diary reports were concordant (e.g., as six of the eight pairs were highly congruent regardless of the symptom being measured - pain, functioning, or anxiety). For those dyads which were discordant, the ratings may have differed based on information used to determine pain (i.e., parents may relay on preconceived expectations; Barakat et al., 2008). Previous studies have also found that lower family income was associated with less concordance in pain reports (Barakat et al., 2006). This was also evident in the present study, as Participants 4 and 5, who were of the lowest socioeconomic status, were highly discordant compared to their caregiver's report on pain reports. It has been suggested that poverty and its associated stressors may reduce accurate communication about pain between caregivers and adolescents (Barakat et al., 2008).

Although it is not possible to identify the specific components of this intervention which contributed to the treatment gains observed (i.e., the "active ingredients"), several likely

candidates are possible. The current treatment intervention included the following components: (a) education, (b) coping strategies, (c) relaxation exercises, (d) cognitive restructuring, (e) goal-setting, (f) problem-solving, and (g) parent-child communication. Although it is possible that changes in any of these factors would be associated with improvements, the components which demonstrated observable change as a result of the intervention included the following: improvements in depressive symptomatology, HRQoL, and total behavior problems (which were statistically detectable effects); reductions in pain and anxiety, and improvements in adaptive coping strategies (which were evident through visual inspection).

Limitations and Future Directions

While there are several benefits to using a single subject design, there are also disadvantages. First, given a multiple-baseline design, statistical evaluation of results was necessarily limited. This, in combination with the small sample size, reduced the power of analyses to detect group differences and to draw definitive conclusions regarding relationships among intervention factors and outcomes. Thus, results in the present study were based primarily on visual inspection which is subject to misinterpretation. Second, in some cases a stable baseline may not have been successfully established (i.e., symptom improvement was observed prior to treatment). Given that a multiple-baseline design depends on changes observed at the onset of intervention, the present results must be interpreted cautiously (Kazdin, 2010).

Third, "inconsistent effects" were observed, in that some of the symptoms changed when the intervention was introduced and others did not, which raises questions about the generality or strength of the intervention (Kazdin, 2010). Fourth, while it is often recommended that the data points from daily diaries be aggregated or averaged over consecutive days or weeks to increase

stability, this data reduction may yield a distorted reflection of daily performance (i.e., lost variability; Kazdin, 2010). Other considerations relevant to this method of assessment include non-adherence to protocol (e.g., participants may fill in the entire diary at one time; Palermo et al., 2003). Furthermore, others have argued that more frequent recordings over the course of the day are needed to track the effects of events on disease symptoms (e.g.,Walker, Garber, Smith, Van Slyke, & Claar, 2001). Future studies might ask youth to keep records at several points during the day such as morning, after school, and bedtime, in order to explore the possibility that there are more immediate effects of potential variables on SCD pain (Walker et al., 2001). Finally, this study did not include a comparison condition other than a staggered baseline.

Although the present study had a successful retention rate, it is important to consider potential difficulties with recruitment and retention of African American families. For the three participants who declined to enter treatment (following the baseline period) potential barriers to treatment may have included socio-demographic factors. These participants were all in the low socioeconomic category (in comparison to the enrolled participants of which only two were in the low socioeconomic status) and experienced transportation challenges (although this was also common to intervention participants). For these participants strategies such as offering incentives for participation and remaining in contact through telephone calls were insufficient to overcome barriers to engagement. Importantly, sociodemographic barriers, mistrust and misunderstanding of the role of research in advancing interventions, and lack of perceived benefits to participation have been shown to contribute to differential drop-out (Barakat, Schwartz, Salamon, & Radcliffe, 2010; Jones, Hadder, Carvajal, Chapman, & Alexander, 2006). Another factor to consider is that the therapist in the present study was of Caucasian ethnicity and although the

extent to which ethnic factors may impact intervention efforts is unclear, this may have been relevant for some participants.

Importantly, this study provides support for the use of a cognitive-behavioral family intervention for children with SCD and the incorporation of culturally sensitive components, (such as the inclusion of the family and African American content). Because the 5-session intervention was time-consuming and given noted transportation difficulties, efforts to develop shorter interventions for pain management, both in clinic and in the homes, may be useful. Although challenges of this intervention study were identified, preliminary data indicate that most participants engaged in and received the intervention well.

Interventions that focus on increasing the coping mechanisms employed by children with SCD and family-focused programs, specifically those that encourage cohesion by emphasizing communication and parent-child interaction, would likely help children and parents deal more efficiently with disease complications (Lutz et al., 2004). The presence of pediatric chronic conditions can be a source of increased stress and distress among family members, alongside other family-specific factors such as socioeconomic status and caregiver marital status. Although family functioning did not show improvements in the present study, it seems likely that successfully targeting this domain would have a collateral impact on successful outcomes. Future studies should continue to explore the variety of factors that are essential to predict children's adjustment to SCD, as increased awareness and knowledge of the variables that affect adjustment to chronic childhood illness can enhance the psychosocial services provided to these families (Lutz et al., 2004).

It will be important for future work to consider cognitive decrements for pediatric patients with SCD. Previous studies have shown impairments in Full Scale, Verbal, and

Performance intelligence quotients for pediatric patients with and without a history of neurologic deficits (Berkelhamm et al., 2007; Puffer, Schatz, & Roberts, 2007). Therefore, it will be important to understand the extent to which a pediatric SCD patient can benefit from the cognitive aspect of CBT therapy (i.e., individual differences in the capacity to conceptualize the relationship between thoughts and their behavior). As an alternative, Acceptance and Commitment Therapy (ACT, Hayes, Strosahl, & Wilson, 1999) may be effective for youth with SCD patients and their families. ACT emphasizes the promotion of daily functioning and quality of life while teaching a willingness to experience difficult and possibly unavoidable private events (e.g., pain, discomfort, fatigue, anxiety) without defense (Hayes, Luoma, Bond, Masuda, & Lillis, 2006). ACT posits that discomfort and pain might be unavoidable and persistent in some contexts, and that experiencing them openly without defense and engaging in value-consistent behaviors are likely to promote greater functioning and quality of life (Masuda, Cohen, Wicksell, Kemani, & Johnson, 2011). Given that preliminary data suggests that ACT can be beneficial for parents as well as adolescents with chronic conditions, a family-based ACT intervention might be particularly useful to children struggling with the management of SCD and should be considered for future studies. Future studies should examine how to best integrate issues of religious coping into clinical conversations (e.g., focusing on coping, adherence, and support) that are practical for the clinician, useful for the family, and will ultimately enhance health outcomes for children with SCD and their families (Cotton et al., 2011). In the present study, religious and existential coping were conceptualized unidimensionally. However, research findings would suggest that both positive and negative aspects of religious coping need to be addressed, and will possibly have different associations with outcomes (Boeving, 2003). As recommended by Schwartz et al. (2007), flexibility in the content of guided imagery and coping

statements is suggested, as culturally congruent and distracting selected images were more accepted than traditional relaxation exercises. Furthermore, positive coping statements that incorporate adolescent language and include reference to spirituality may be most effective for African Americans (Schwartz et al., 2007).

It will also be important for future work to clarify the linkages among pain, coping, and internalizing symptoms, given the greater reported pain and increased risk for symptoms of depression and anxiety in adolescence (Barakat et al., 2007; Dampier, Ely, Brodecki, & O'Neal, 2002). In addition, it is critical to examine factors that may be relevant for developmentally appropriate modifications to intervention. For example, developing effective intervention strategies to support transitions in independence (from childhood to adolescence, and adolescence to adulthood) seems relevant for effectively enhancing optimal adaptation and disease management (Barakat et al., 2007). Finally, it will be important to consider variability in pain symptoms for pediatric SCD. It is often the case that more severe sickle cell pain is treated with medication such as hydroxyurea. However, it is unclear how medication in conjunction with CBT, or medication alone, may differentially impact treatment outcome. Therefore, it will be important for future research to address these questions.

Overall, the present study provides preliminary support for the utility of family-based and culturally sensitive cognitive-behavioral intervention for pediatric SCD. Although there were important limitations noted, results are promising, in that several domains of change and improvement were associated with the intervention.

References

Achenbach, T.M., & Edelbrock, C. (1983). *Manual for the Child Behavior Checklist and Revised Child Behavior Profile.* Burlington: University of Vermont, Department of Psychiatry.

Agency for Health Care Policy and Research. (1993). *Sickle cell disease: Screening, diagnosis, management, and counseling in newborns and infants.* Rockville, MD: Department of Health and Human Services.

Amrolia, P.J., Almeida, A., Davies, S.C., & Roberts, I.A. (2003). Therapeutic challenges in childhood sickle cell disease. Part 2: A problem-oriented approach. *British Journal of Haematology, 120*, 737-743.

Arnstein, P., Caudill, M., Mandle, C.L., Norris, A., & Beasley, R. (1999). Self-efficacy as a mediator of the relationship between pain intensity, disability and depression in chronic pain patients. *Pain, 80*, 483-491.

Ballas, S.K. (2002). Sickle cell anemia: Progress in pathogenesis and treatment. *Drugs, 62*, 1143-1172.

Bandura, A. (1988). Self-efficacy: Toward a unifying theory of behavioral change. *Psychological Review, 84,* 191-215.

Barbarin, O.A., & Christian, M. (1999). The social and cultural context of coping with sickle cell disease: I. A review of biomedical and social issues. *Journal of Black Psychology, 25*, 277-293.

Barbarin, O.A., Whitten, C.F., & Bond, S. (1994). Estimating rates of psychosocial problems in urban and poor children with sickle cell anemia. *Health and Social Work, 19*, 112-119.

Barakat, L.P., Schwartz, L.A., Salamon, K.S., & Radcliffe, J. (2010). A family-based randomized controlled trial of pain intervention for adolescents with Sickle Cell Disease. *Journal of Pediatric Hematology/Oncology, 32,* 540-547.

Barakat, L.P., Simon, K., Schwartz, L.A., Radcliffe, J. (2008). Correlates of pain-rating concordance for adolescents with Sickle Cell Disease and their caregiver. *The Clinical Journal of Pain, 24(5),* 438-446.

Barakat, L.P., Schwartz, L.A., Simon, K., & Radcliffe, J. (2007). Negative thinking as a coping strategy mediator of pain and internalizing symptoms in adolescents with Sickle Cell Disease. *Journal of Behavioral Medicine, 30,* 199-208.

Barakat, L.P., Lash, L.A., Lutz, M.J., & Nicolaou, D.C. (2006). Psychosocial adaptation of children and adolescents with sickle cell disease. In R.T. Brown (Ed.), *Comprehensive handbook of childhood cancer and sickle cell disease: A biopsychosocial approach* (pp. 471-495). New York: Oxford University Press.

Barakat, L.P., Lutz, M., Smith-Whitley, K., & Ohene-Frempong, K. (2005). Is treatment adherence associated with better quality of life in children with Sickle Cell Disease? *Quality of Life Research, 14,* 407-414.

Barakat, L.P., Smith-Whitley, K., & Ohene-Frempong, K. (2002). Treatment adherence in children with sickle cell disease: Disease-related risk and psychosocial resistance factors. *Journal of Clinical Psychology in Medical Settings, 9,* 201-205.

Baskin, M.L., Collins, M.H., Brown, F., Griffith, J.R., Samuels, D., Moody, A. et al. (1998). Psychosocial considerations in sickle cell disease (SCD): The transition from adolescence to young adulthood. *Journal of Clinical Psychology in Medical Settings, 5,* 315-341.

Benjamin, L.J., Dampier, C.D., Jacox, A.K., Odesina, V., Phoenix, D., Shapiro, B. et al. (1999). *Guideline for the management of acute and chronic pain in sickle cell disease* (APS Clinical Practice Guidelines Series, No. 1). Glenview, IL: American Pain Society.

Berkelhamm, L.D., et al. (2007). Neurocognitive sequelae of pediatric Sickle Cell Disease: A review of the literature. *Child Neuropsychology, 13,* 120-131.

Beyer, J., Simmons, L., Woods, G., & Woods, P. (1999). A chronology of pain and comfort in children with sickle cell disease. *Archives of Pediatric and Adolescent Medicine, 153*, 913-920.

Britt, G. C. (1995). *Children's coping with everyday stressful situations: The role played by religion.* Poster presented at the 61st Biennial Meeting of the Society for Research in Child Development, Indianapolis, Indiana.

Boeving, C.A. (2003). *Coping and Adjustment in Child Cancer Survivors: An Investigation into Spirituality as a Predictor of Psychosocial Outcomes* . Unpublished doctoral dissertation, Virginia Polytechnic Institute and State University, Virginia.

Boyd-Franklin, N. (2003). *Black families in therapy: Understanding the African American experience* (3rd ed.). New York: Guilford Press.

Bradley, L.A. (1996). In: R.J. Gatchel, D.C., Turk (Eds.), *Psychological Approaches to Pain Management: A Practitioner's Handbook*. New York: The Guilford Press, pp. 131-147.

Bronfenbrenner, U. (1977). Ecology of the family as a context for human development: Research perspectives. *Developmental Psychology*, *22,* 723–742.

Brown, R.T., Lambert, R., Devine, D., Baldwin, K., Casey, R., Doepke, K., et al. (2000). Risk-resistance adaptation model for caregivers and their children with sickle cell syndromes. *Annals of Behavioral Medicine, 22,* 158-169.

Brown, R.T., Eckman, J., Baldwin, K., Buchanan, I., & Dingle, A.D. (1995). Prospective aspects of adaptive behavior in children with sickle cell syndromes. *Children's Health Care, 24,* 205-222.

Brown, R.T., Kaslow, N.J., Doepke, K., Buchanan, I., Eckman, J., Baldwin, K., et al. (1993). Psychosocial and family functioning in children with sickle cell syndrome, and their mothers. *Journal of the American Academy of Child and Adolescent Psychiatry, 32,* 545-553.

Burlew, K. (2002). Empirically derived guidelines for assessing the psychosocial needs of children and adolescents with sickle cell. *Social Work in Health Care, 36,* 29-44.

Burlew, K., Telfair, J., Colangelo, L., & Wright, W.C. (2000). Factors that influence adolescent adaptation to sickle cell disease. *Journal of Pediatric Psychology, 25,* 287-299.

Burlew, A.K., Evans, R., & Oler, C. (1989). The impact of a child with sickle cell disease on family dynamics. *Annals of the New York Academy of Sciences, 565,* 161-171.

Carr, J. (2005). Recommendations for reporting multiple-baseline designs across participants. *Behavioral Interventions, 20*(3), 219-224.

Casey, R.L., Brown, R.T., & Bakeman, R. (2000). Predicting adjustment in children and adolescents with sickle cell disease: A test of the risk-resistance—adaptation model. *Rehabilitation Psychology, 45*, 155-178.

Chambers, C.T. (2003). The role of family factors in pediatric pain. In P.J. McGrath, G.A. Finley (Eds.), *Pediatric pain: Biological and social context* (pp. 99-130). Seattle, WA: IASP Press.

Charache, S., Lubin, B., & Reid, C.D. (1989). *Management and Therapy of Sickle Cell Disease*, NIH publication No. 89-2117, National Institutes of Health, Washington DC.

Chen, E., Cole, S.W., & Kato, P.M. (2004). A review of empirically supported psychosocial interventions for pain and adherence outcomes in sickle cell disease. *Journal of Pediatric Psychology, 29*, 197-209.

Cotton, S., Grossosehme, D., & McGrady, M.E. (2011). Religious coping and the use of prayer in children with Sickle Cell Disease. *Pediatric Blood & Cancer.* [Epub ahead of print]

Coughlan, G.M., Ridout, K.L., Williams, A., & Richardson, P.H. (1995). Attrition from a pain management program. *British Journal of Clinical Psychology, 343*, 471-479.

Dampier, C., Ely, B., Brodecki, D., & O'Neal, P. (2002). Characteristics of pain management at home in children and adolescents with Sickle Cell Disease by using dairy self-reports. *The Journal of Pain, 3(6),* 461-470.

Department of Health and Human Services. *Healthy People 2020.* Washington, D.C.: January 2011. Available on-line: http://www.healthypeople.gov/2020

Drotar, D. (1997). Relating parent and family functioning to the psychological adjustment of children with chronic health conditions: What have we learned? What do we need to know? *Journal of Pediatric Psychology, 22,* 149-165.

Dunn-Grier, B., McGrath, P.J., Rourke, B.P., Latter, J., & D'Astous, J. (1986). Adolescent chronic pain: The ability to cope. *Pain, 26,* 23-32.

Dworkin, R.H. et al. (2008). Consensus Statement: Interpreting the clinical importance of treatment outcomes in chronic pain clinical trials: IMMPACT Recommendations. *The Journal of Pain, 9(2),* 105-121.

Elander, J., & Midence, K. (1996). A review of evidence about factors affecting quality of pain management in sickle cell disease. *The Clinical Journal of Pain, 12*, 180-193.

Fuggle, P., Shand, P.A.X., Gill, L.J., & Davies, S.C. (1996). Pain, quality of life and coping in sickle cell disease. *Archives of Disease in Childhood, 75,* 199-203.

Gadbury, G.L., Coffey, C.S., & Allison, D.B. (2003). Modern statistical methods for handling missing repeated measurements in obesity trial data: Beyond LOCF. *Obesity Reviews, 4,* 175-184.

Gil, K.M., Carson, J.W., Porter, L.S., Ready, J., Valrie, C., Redding-Lallinger, R., et al. (2003). Daily stress and mood and their association with pain, health-care use, and school activity in adolescents with sickle cell disease. *Journal of Pediatric Psychology, 28,* 363-373.

Gil, K.M., Anthony, K.K., Carson, J.W., Redding-Lallinger, R., Daeschner, C.W., & Ware, R.E. (2001). Daily coping practice predicts treatment effects in children with sickle cell disease. *Journal of Pediatric Psychology, 26,* 163-173.

Gil, K.M., Porter, L., Ready, J., Workman, E., Sedway, J., & Anthony, K.K. (2000). Pain in children and adolescents with Sickle Cell Disease: An analysis of daily pain diaries. *Children's Health Care, 29(4),* 225-241.

Gil, K.M., Wilson, J.J., Edens, J.L., Workman, E., Ready, J., Sedway, J., Redding-Lallinger, R., & Daeschner, C.W. (1997). Cognitive coping skills training in children with sickle cell disease pain. *International Journal of Behavioral Medicine, 4*, 364-377.

Gil, K.M., Thompson, R.J., Keith, B.R., Tota-Faucetter, M., Noll, S., & Kinney, T.R. (1993). Sickle cell disease pain in children and adolescents: Change in pain frequency and coping strategies over time. *Journal of Pediatric Psychology, 18,* 621-637.

Gil, K.M., Williams, D.A., Thompson, R.J., & Kinney, T.R. (1991). Sickle cell disease in children and adolescents: The relation of child and parent pain coping strategies to adjustment. *Journal of Pediatric Psychology, 16,* 643-663.

Gil, K.M., Abrams, M.R., Phillips, G., & Keefe, F.J. (1989). Sickle cell disease pain: Relation of coping strategies to adjustment. *Journal of Consulting and Clinical Psychology, 57*, 725-731.

Green, B.L., Patridge, E.E., Fouad, M.N., Kohler, C., Crayton, E.F., & Alexander, L. (2000). African-American attitudes regarding cancer clinical trials and research studies: Results from focus group methodology. *Ethnicity & Disease, 10,* 76-86.

Grootenhuis, M.A., Koopman, H.M., Verrips, E.G.H., Vogels, A.G.C., & Last, B.F. (2007). Health-related quality of life problems of children aged 8-11 years with a chronic disease. *Developmental Neurorehabilitation, 10(1),* 27-33.

Harvey, M.T., May, M.E., & Kennedy, C.H. (2004). Nonconcurrent Multiple Baseline Designs and the Evaluation of Educational Systems. *Journal of Behavioral Education, 13,* 267-276.

Hayes, S.C., Strosahl, K.D., & Wilson, K.G. (1999). *Acceptance and Commitment Therapy: An exceptional approach to behavior change.* New York: Guilford Press.

Hayes, S.C., Luoma, J.B., Bond, F.W., Masuda, A., & Lillis, J. (2006). Acceptance and commitment therapy: Model, processes, and outcome. *Behavior Research and Therapy, 44,* 1-25.

Haythornthwaite, J.A., Menefee, L.A., Heinberg, L.J., & Clark, M.R. (1998). Pain coping strategies predict perceived control over pain. *Pain, 77*, 33-39.

Hocking, M.C., & Lochman, J.E. (2005). Applying the transactional stress and coping model to sickle cell disorder and insulin-dependent diabetes mellitus: Identifying psychosocial variables related to adjustment and intervention. *Clinical Child and Family Psychology Review, 8*, 221-246.

Hoppe, C., Styles, L., & Vichinsky, E. (1998). The natural history of sickle cell disease. *Current Opinion in Pediatrics, 10*, 49-52.

Hunfeld, J.A.M., Perquin, C.W., Duivenvoorden, H.J., Hazenbroek-Kampschreur, A.A.J.M., Passchier, J., van Suijlekom-Smit, L.W.A., et al. (2001). Chronic pain and its impact on quality of life in adolescents and their families. *Journal of Pediatric Psychology, 26,* 145-153.

Hurtig, A.L. (1994). Relationships in families of children and adolescents with sickle cell disease. *Journal of Health and Social Policy, 5,* 161-183.

Hurtig, A.L., Koepke, D., & Park, K.B. (1989). Relation between severity of chronic illness and adaptation in children and adolescents with sickle cell disease. *Journal of Pediatric Psychology, 14,* 117-132.

Hurtig, A.L., & White, L.S. (1986). Psychological adjustment in children and adolescents with sickle cell disease. *Journal of Pediatric Psychology, 11*, 411-427.

Ievers-Landis, C.E., Brown, R.T., Drotar, D., Bunke, V., Lambert, R.G., & Walker, A.A. (2001). Situational analysis of parenting problems for caregivers of children with sickle cell syndromes. *Journal of Developmental and Behavioral Pediatrics, 22,* 169-178.

Jacob, E., Miaskowski, C., Savedra, M., Beyer, J.E., Treadwell, M., & Styles, L. (2003). Management of vaso-occlusive pain in children with sickle cell disease. *Journal of Pediatric Hematology/Oncology, 25*, 307-311.

Jensen, M.P., Turner, J.A., Romano, J.M., & Karoly, P. (1991). Coping with chronic pain: A critical review of the literature. *Pain, 47,* 249-283.

Johnston, J.M. & Pennypacker, H.S. (1993). Strategies and tactics of behavioral research (2nd ed.). Hillsdale, NJ, England: Lawrence Erlbaum Associates, Inc.

Jones, R.T., Hadder, J., Carvajal, F., Chapman, S., & Alexander, A. (2006). Conducting research in diverse, minority, and marginalized communities. In F. Norris, S. Galea, M. Friedman, & P. Watson (Eds), *Methods of Disaster Mental Health Research* (pp. 265-277). New York: Guilford Press.

Kaslow, N.J., Collins, M.H., Rashid, F.L., Baskin, M.L., Griffith, J.R., Hollins, L., et al. (2000). The efficacy of a pilot family psychoeducational intervention for pediatric sickle cell disease (SCD). *Families, Systems, and Health, 8,* 381-404.

Kaslow, N.J., & Brown, F. (1995). Culturally sensitive family interventions for chronically ill youth: Sickle cell disease as an example. *Family Systems Medicine, 13,* 201-213.

Kazdin, A.E. (2010). *Single-case research designs: Methods for clinical and applied settings* (2nd ed.). New York: Oxford University Press.

Kazdin, A.E. & Weisz, J.R. (1998). Identifying and developing empirically supported child and adolescent treatments. *Journal of Consulting and Clinical Psychology, 66*(1), 19-36.

Kell, R.S., Kliewer, W., Erickson, M.T., & Ohene-Frempong, K. (1998). Psychological adjustment of adolescents with sickle cell disease: Relations with demographic, medical, and family competence variables. *Journal of Pediatric Psychology, 23*, 301-312.

Kliewer, W., & Lewis, H. (1995). Family influences on coping processes in children and adolescents with sickle cell disease. *Journal of Pediatric Psychology, 20,* 511-525.

Kovacs, M., & Beck, A.T. (1977). An empirical-clinical approach toward a definition of children depression. In J.G. Schulterbrandt & A. Raskin (Eds.), *Depression in childhood: Diagnosis, treatment, and conceptual models.* New York: Raven.

La Greca, A.M., & Bearman, K.J. (2001). Commentary: If 'An apple a day keeps the doctor away,' why is adherence so hard? *Journal of Pediatric Psychology, 26,* 279-282.

Landolt, M. A., Vollrath, M., & Ribi, K. (2002). Predictors of coping strategy selection in pediatric patients. *Acta Paediatrica, 91,* 954-960.

Lazarus, R.S., & Folkman, S. (1984). *Stress, appraisal, and coping.* New York: Springer.

Lemanek, K.L., Moore, S.L., Gresham, F.M., Williamson, D.A., & Kelley, M.L. (1986). Psychological adjustment of children with sickle cell anemia. *Journal of Pediatric Psychology, 11,* 397-410.

Lewis, H.A., & Kliewer, W. (1996). Hope, coping, and adjustment among children with sickle cell disease: Tests of mediator and moderator models. *Journal of Pediatric Psychology, 21,* 25-41.

Lutz, M.J., Barakat, L.P., Smith-Whitley, K., & Ohene-Frempong, K. (2004). Psychological children with sickle cell disease: Family functioning and coping. *Rehabilitation Psychology, 49,* 224-232.

Mallinckrodt, C.H., et al. (2003). Assessing response profiles from incomplete longitudinal clinical trial data under regulatory considerations. *Journal of Biopharmaceutical Statistics, 13,* 179-190.

Martinez, C. R., & Eddy, J. M. (2005). Effects of culturally adapted parent management training on Latino youth behavioral health outcomes. Journal of Consulting and Clinical Psychology, 73, 841–851.

Masuda, A., Cohen, L.L., Wicksell, R.K., Kemani, M.K., & Johnson, A. (2011). A case study: Acceptance and commitment therapy for pediatric Sickle Cell Disease. *Journal of Pediatric Psychology, 35,* 1-11.

Merlijn, V.P.B.M., Hunfeld, J.A.M., van der Wouden, J.C., Hazebroek-Kampschreur, A.A.J.M., van Suijlekom-Smit, L.W.A., Koes, B.W. et al. (2005). A cognitive-behavioural program for adolescents with chronic pain—a pilot study. *Patient Education and Counseling, 59,* 126-134.

Merskey, H., & Bogduk, N. (Eds.). (1994). *Classification of chronic pain: Descriptions of chronic pain syndromes and definitions of pain terms* (2nd ed.). Seattle, WA: IASP Press.

Midence, K., McManus, C., Fuggle, P., & Davies, S. (1996). Psychological adjustment and family functioning in a group of British children with sickle cell disease: Preliminary empirical findings and meta-analysis. *British Journal of Clinical Psychology, 35,* 439-450.

Morley, S., Eccleston, C., & Williams, A.C. (1999). Systematic review and meta-analysis of randomized controlled trials of cognitive behavior therapy for chronic pain in adults, excluding headaches. *Pain, 80,* 1-13.

Moos, R.H., & Moos, B.S. (1981). *Family Environment Scale manual.* Palo Alto, CA: Consulting Psychologists Press.

Murray, N., & May, A. (1988). Painful crises in sickle cell disease. *British Medical Journal, 297*, 452-454.

Murry, V.M., Bynum, M.S., Brody, G.H., Willert, A., & Stephens, D. (2001). African American single mothers and children in context: A review of studies on risk and resilience. *Clinical Child and Family Psychology Review, 4,* 133-155.

Nevergold, B.S. (1987). Therapy with families of children with sickle cell disease. *Family Therapy Collection, 22,* 67-79.

Noll, R.B., Swecki, E., Garstein, M., Vannatta, K., Kalinyak, K., Davies, W.H. et al. (1994). Parental distress, family conflict, and role of social support for caregivers with or without a child with sickle cell disease. *Family Systems Medicine, 12,* 281-294.

Noll, R.B., Vannatta, K., Koonz, K., Kalinyak, K., Bukowski, W.M., & Davies, W.H. (1986). Peer relationships and emotional well-being of youngsters with sickle cell disease. *Child Development, 67,* 423-436.

Palermo, T.M., Witherspoon, D., Valenzuela, D., & Drotar, D. (2004). Development and validation of the Child Activity Limitations Interview: A measure of pain-related functional impairment in school-age children and adolescents. *Pain, 109*, 461-470.

Palermo, T.M., Valenzuela, D., & Stork, P.L. (2003). A randomized trial of electronic versus paper pain diaries in children: Impact on compliance, accuracy, and acceptability. *Pain, 107,* 213-219.

Palermo, T.M., Schwartz, L., Drotar, D., & McGowan, K. (2002). Parental report of health related quality of life in children with sickle cell disease. *Journal of Behavioral Medicine, 25*, 269-283.

Palermo, T.M. (2000). Impact of recurrent and chronic pain on child and family daily functioning: A critical review of the literature. *Developmental and Behavioral Pediatrics, 21*, 2000.

Pallister, C.J. (1992). A crisis that can be overcome: Management of sickle cell disease. *Professional Nurse, 2,* 509-513.

Petermann, F., & Muller, J.M. (2001). *Clinical Psychology and Single-Case Evidence: Practical Approach to Treatment, Planning, and Evaluation.* New York: John Wiley & Sons Ltd.

Petersen, C.C., & Palermo, T.M. (2004). Parental reinforcement of recurrent pain: The moderating impact of child depression and anxiety on functional disability. *Journal of Pediatric Psychology, 29(5),* 331-341.

Pfefferbaum, B., Adams, J., & Aceves, J. (1990). The influence of culture on pain in Anglo and Hispanic children with cancer. *Journal of the American Academy of Child and Adolescent Psychiatry, 29,* 642-647.

Platt, O.S., Thorington, B.D., Brambilla, D.J., Milner, P.F., Rosse, W.F., Vichinsky, E., & Kinney, T.R. (1991). Pain in sickle cell disease: Rates and risk factors. *New England Journal of Medicine, 325*, 11-16.

Powers, S.W., Jones, J.S., & Jones, B.A. (2005). Behavioral and cognitive-behavioral interventions with pediatric populations. *Clinical Child Psychology and Psychiatry, 10*, 65-77.

Powers, S.W., Mitchell, M.J., Graumlich, S.E., Byars, K.C., & Kalinyak, K.A. (2002). Longitudinal assessment of pain, coping, and daily functioning in children with sickle cell disease receiving pain management skills training. *Journal of Clinical Psychology in Medical Settings, 9,* 109-119.

Puffer, E. Schatz, J., & Roberts, C.W. (2007). The association of oral hydroxyurea therapy with improved cognitive functioning in Sickle Cell Disease. *Child Neuropsychology, 13,* 142-154.

Radcliffe, J., Barakat, L.P., & Boyd, R.C. (2006). Family systems issues in pediatric sickle cell disease. In R.T. Brown (Ed.), *Comprehensive handbook of childhood cancer and sickle cell disease: A biopsychosocial approach* (pp. 496-513). New York: Oxford University Press.

Raghupathy, R., & Billett, H.H. (2009). Promising therapies in sickle cell disease. *Cardiovascular & Haematological Disorders-Drug Targets, 9,* 1-8.

Reid, G.J., Gilbert, C.A., & McGrath, P.J. (1998). The Pain Coping Questionnaire: Preliminary validation. *Pain, 76,* 83-96.

Reynolds, C., & Richmond, B.O. (1985). *Revised Children's Manifest Anxiety Scale.* Los Angeles: Western Psychological Services.

Richards, S. B., Taylor, S. B., Ramasamy, R., & Richards, R. Y. (1999). Single-subject research. Applications in educational and clinical settings. San Diego, CA: Singular.

Robins, P.M., Smith, S.M., Glutting, J.J., & Bishop, C.T. (2005). A randomized controlled trial of a cognitive-behavioral family intervention for pediatric recurrent abdominal pain. *Journal of Pediatric Psychology, 30*, 397-408.

Sanders, M.R., Shepherd, R.W., Cleghorn, G., & Woolford, H. (1994). The treatment of recurrent abdominal pain in children: A controlled comparison of cognitive-behavioral family intervention and standard pediatric care. *Journal of Consulting and Clinical Psychology, 62*, 306-314.

Schwartz, L.A., Radcliffe, J., & Barakat, L.P. (2007). The development of a culturally sensitive pediatric pain management intervention for African American adolescents with sickle cell disease. *Children's Healthcare, 36*, 267-283.

Scott, K.D., & Scott, A.A. (1999). Cultural therapeutic awareness and sickle cell anemia. *Journal of Black Psychology, 25,* 316-335.

Shapiro, B.S., Dinges, D.F., Orne, E.C., Bauer, N., Reilly, L.B., Whitehouse, W.G., Ohene-Frempong, K., & Orne, M. (1995). Home management of sickle cell-related pain in children and adolescents: Natural history and impact on school attendance. *Pain, 61*, 139-144.

Shapiro, B. (1993). Management of painful episodes in sickle cell disease. In N. Schechter, C. Berde, & M. Yaster (Eds.), *Pain in infants, children, and adolescents* (pp. 385-410). Baltimore: Williams & Wilkins.

Shapiro, B.S., Dinges, D.F., Orne, E.C., Ohene-Frempong, K., & Orne, M.T. (1990). Recording of crisis pain in sickle cell disease. *Advances in Pain Research Therapy, 15*, 313-321.

Sharpe, J.N., Brown, R.T., Thompson, N.J., & Eckman, J. (1994). Predictors of coping with pain in mothers and their children with sickle cell syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry, 33,* 1246-1255.

Spirito, A., Stark, L.J., Gil, K.M., & Tyc, V.L. (1995). Coping with everyday and disease-related stressors by chronically ill children and adolescents. *Journal of the American Academy of Child and Adolescent Psychiatry, 34,* 283-290.

Steinberg, M.H. (1999). Management of sickle cell disease. *New England Journal of Medicine, 340*, 1021-1029.

Stresisand, R., Braniecki, S., Tercyak, K.P., & Kazak, A.E. (2001). Childhood illness-related parenting stress: The pediatric inventory for parents. *Journal of Pediatric Psychology, 26,* 155-162.

Strickland, B.R. (1978). Internal-external expectancies and health-related behaviors. *Journal of Consulting and Clinical Psychology, 46,* 1192-1211.

Swain, N.F., Mitchell, M.J., & Powers, S.W. (2006). Pain management of sickle cell disease. In R.T. Brown (Ed.), *Comprehensive handbook of childhood cancer and sickle cell disease: A biopsychosocial approach* (pp. 514-532). New York: Oxford University Press.

Tarnowski, K., & Brown, R.T. (2000). Psychological aspects of pediatric disorders. In M. Hersen & R. Ammerman (Eds.), *Advanced Abnormal Child Psychology* (2nd ed., pp. 131-152). Hillsdale, NJ: Erlbaum.

Taylor, R.D, & Roberts, D. (1995). Kinship support and maternal and adolescent well-being in economically disadvantaged African American families. *Child Development, 66,* 1585-1597.

Telfair, J., & Gardner, M.M. (1999). African American adolescents with sickle cell disease: Support groups and psychological well-being. *Journal of Black Psychology, 25*, 378-390.

Thompson, R.J., Armstrong, F.D., Link, C.L., Pegelow, C.H., Moser, F., & Wang, W.C. (2003). A prospective study of the relationship over time of behavior problems, intellectual functioning, and family functioning in children with sickle cell disease: A report from the Cooperative Study of Sickle Cell Disease. *Journal of Pediatric Psychology, 28,* 59-65.

Thompson, R.J., Gustafson, K.E., Gil, K.M., Godfrey, J., & Bennet-Murphy, L.M. (1998). Illness specific patterns of psychological adjustment and cognitive adaptational process in children with cystic fibrosis and sickle cell disease. *Journal of Clinical Psychology, 54,* 121-128.

Thompson, R.J., & Gustafson, K.E. (1996). *Adaptation to chronic childhood illness.* Washington, DC: American Psychological Association Press.

Thompson, R.J., Gustafson, K., & Gil, K.M. (1995). Psychological adjustment of adolescents with cystic fibrosis or sickle cell disease and their mothers. In J. Wallander & L. Siegal (Eds.), *Advances in pediatric psychology: II. Behavioral perspectives on adolescent health* (pp. 232-247). New York: Guilford.

Thompson, R.J., Gil, K.M., Keith, B.R., Gustafson, K.E., George, L.K., & Kinney, T.R. (1994). Psychological adjustment of children with sickle cell disease: Stability and change over a 10-month period. *Journal of Consulting and Clinical Psychology, 62*, 856-860.

Thompson, R.J., Gustafson, K.E., George, L.K., & Spock, A. (1994). Change over a 12-month period in the psychosocial adjustment of children and adolescents with cystic fibrosis. *Journal of Pediatric Psychology, 19*, 189-203.

Thompson, R.J., Gil, K.M., Burbach, D.J., Keith, B.R., & Kinney, T.R. (1993a). Role of child and maternal processes in the psychological adjustment of children with sickle cell disease. *Journal of Consulting and Clinical Psychology, 61*, 468-474.

Thompson, R.J., Gil., K.M., Burbach, D.J., Keith, B.R., & Kinney, T.R. (1993b). Psychological adaptation of mothers of children and adolescents with sickle cell disease: The role of stress, coping methods, and family functioning. *Journal of Pediatric Psychology, 18,* 549-559.

Thompson, R.J., Jr., Gustafon, K.E., Hamlett, K.W., & Spock, A. (1992). Stress, coping, and family functioning in the psychological adjustment of mothers of children and adolescents with cystic fibrosis. *Journal of Pediatric Psychology, 17,* 741-755.

Turk, D.C., & Okifuji, A. (2001). Pain terms and taxonomies of pain. In J.D. Loeser, S.H. Butler, C.R. Chapman, & D.C. Turk (Eds.), *Bonica's management of pain* (3rd ed., pp. 17-25). Philadelphia: Lippincott, Williams and Wilkins.

Turner, J.A., & Jensen, M.P. (1993). Efficacy of cognitive therapy for chronic low back pain. *Pain, 52,* 169-177.

Unnebrink, K., & Windeler, J. (2001). Intention-to-treat: Methods for dealing with missing values in clinical trials of progressively deteriorating diseases. *Statistics in Medicine, 20,* 3931-46.

Van Slyke, D.A., & Walker, L.S. (2006). Mothers' responses to adolescent's pain. *Clinical Journal of Pain, 22*, 387-391.

Varni, J.W., Seid, M., & Rode, C.A. (1999). The PedsQL: Measurement model for the pediatric quality of life inventory. *Medical Care, 37,* 126-139.

Varni, J.W., Thompson, K.L., & Hanson, V. (1987). The Varni/Thompson Pediatric Pain Questionnaire. I. Chronic musculoskeletal pain in juvenile rheumatoid arthritis. *Pain, 28*, 27-38.

Vervoot, T., Goubert, L., Eccleston, C., Bijttebier, P., & Crombez, G. (2006). Catastrophic thinking about pain is independently associated with pain severity, disability, and somatic complaints in school children and children with chronic pain. *Journal of Pediatric Psychology, 31(7),* 674-683.

Walker, L.S., Smith, C.A., Garber, J., & Claar, R.L. (2005). Testing a model of pain appraisal and coping in adolescent with chronic abdominal pain. *Health Psychology, 24,* 364-374.

Walker, L. S., Garber, J., Smith, C. A., Van Slyke, D. A., & Claar, R. L. (2001). The relation of daily stressors to somatic and emotional symptoms in children with and without recurrent abdominal pain. *Journal of Consulting and Clinical Psychology, 69,* 85–91.

Walker, L.S., Smith, C.A., Garber, J., & Van Slyke, D.A. (1997). Development and validation of the pain response inventory for adolescent. *Psychological Assessment, 9,* 392-405.

Walker, L.S., & Greene, J.W. (1991). Negative life events and symptom resolution in pediatric abdominal pain patients. *Journal of Pediatric Psychology, 16*, 341-360.

Walker, D.K., & Jacobs, F.H. (1984). Chronically ill children in school. *Peabody Journal of Education, 61*, 29-71.

Wallander, J.L., & Thompson, R.J. (1995). Psychosocial adjustment of children with chronic physical conditions. In M.C. Roberts (Ed.), *Handbook of Pediatric Psychology* (2nd ed., pp. 124-141). New York: Guilford Press.

Wallander, J.L., & Varni, J. (1992). Adjustment in children with chronic physical disorders: Programmatic research on a disability-stress-coping model. In A.M. LaGreca, L.J. Siegel, J.L. Wallander, & C.E. Walker (Eds.), *Stress and Coping in Child Health* (pp. 279-297). New York: Guilford Press.

Wilson, M.N. (1989). Child development in the context of the black extended family. *American Psychologist, 44,* 380-385.

Zehnder, D., Prchal, A., Vollrath, M., & Landolt, M.A. (2006). Prospective study of the effectiveness of coping in pediatric patients. *Child Psychiatry and Human Development, 36,* 351-368.

Table 1. Participant Demographics

Participant	Sex	Age	Diagnosis	Parent Education	Parent Marital Status	Annual Income
1	Male	14	Hb SS	College Graduate	Married	$35,000-$50,000
2	Female	13	Hb SS	College Graduate	Married	$35,000-$50,000
3	Male	8	Hb SC	Some College	Married	$65,000-$80,000
4	Male	13	Hb SS	High School Graduate	Never Married	$10,000-$20,000
5	Female	9	Hb SS			
6	Female	15	Hb SS	Some College	Widowed	$65,000-$80,000
7	Male	12	Hb SC	College Graduate	Divorced	$20,000-$30,000
8	Female	11	Hb SS	College Graduate	Never Married	$20,000-$30,000

Table 2. Parent-Child Concordance for Daily Diary Ratings

Participant	Correlations		
	Pain	Functionality	Anxiety
1	.869** (.000)	.922** (.000)	.231* (.05)
2	.753** (.000)	1.00** (.000)	.415** (.000)
3	.826** (.000)	1.00** (.000)	1.00** (.000)
4	-.035 (.8)	.999** (.000)	-.472** (.000)
5	.092 (.5)	1.00** (.000)	-.107 (.5)
6	.768** (.000)	.999** (.000)	.323** (.01)
7	.849** (.000)	.140 (.2)	.787** (.000)
8	.300** (.01)	-.122 (.3)	-.026 (.8)

** indicates $p<.001$

Table 3. Mean Weekly Pain Intensity (Youth Report) on Visual Analog Scale

Participant	Mean Pain Intensity*		Treatment Change Score†
	Baseline	Treatment	
1	3	3.1	+ .1
2	1.1	1	**- .1**
3	2	1	**- 1**
4	3	1	**- 2**
5	1	1.2	+ .2
6	3	1.3	**- 2.7**
7	6	2	**- 4**
8	1	2	+ 1

*Mean Pain Intensity was calculated on days with pain using visual analogue scale (range from 0 = no pain to 10 = severe pain).

†Treatment change scores were calculated (Treatment – Baseline). Negative (bold) values represent a mean pain intensity decrease from baseline to intervention.

Table 4. Mean Weekly Functionality for Baseline and Treatment Phases (Youth Report)

Participant	Mean Weekly Functionality*		Treatment Change Score†
	Baseline	Treatment	
1	.5	2	+1.5
2	1.3	.6	**-.7**
3	1.4	1.2	**-.2**
4	2.2	1.8	**-.4**
5	1.3	1.4	+.1
6	1.5	1.9	+.4
7	2.1	2.2	+.1
8	1.3	6	**-.7**

*Range = 0 (no activity reduction) to 5 (high reduction of activity)

†Treatment change scores were calculated (Treatment – Baseline). Negative (bold) values represent a mean pain intensity decrease from baseline to intervention.

Table 5. Youth Report of Mean Weekly Anxiety for Baseline and Treatment Phases

Participant	Mean Weekly Anxiety*		Treatment Change Score†
	Baseline	Treatment	
1	8.5	7	**-1.5**
2	0	0	0
3	5.3	4	**-1.3**
4	8	8	0
5	1	4	+3
6	8	8	0
7	15	20	+5
8	12	12	0

*Range = 0 (no anxiety) to 20 (high anxiety)

†Treatment change scores were calculated (Treatment – Baseline). Negative (bold) values represent a mean pain intensity decrease from baseline to intervention.

Table 6. Functional Disability Inventory Total Scores

Participant	Measure (Total Score)	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	Child	0	21	7	0	19
	Parent	0	22	21	0	19
2	Child	13	3	5	8	8
	Parent	7	2	0	3	5
3	Child	0	9	4	1	2
	Parent	43	0	2	0	0
4	Child	11	2	0	13	7
	Parent	2	0	6	0	2
5	Child	8	0	0	0	0
	Parent	2	1	0	0	0
6	Child	5	8	10	-	-
	Parent	0	7	3	-	-
7	Child	2	17	3	0	17
	Parent	4	6	3	8	19
8	Child	9	6	-	-	-
	Parent	3	2	-	-	-
Overall Means	Child	6.00 (5.01)	8.25 (7.36)	4.14 (3.63)	3.67 (5.54)	10.20 (7.79)
	Parent	7.63 (14.47)	5.00 (7.35)	5.00 (7.35)	1.83 (3.25)	9.00 (9.30)

Higher scores indicate greater perceived functional disability

Table 7. Pediatric Quality of Life Inventory Total HRQoL Scores

Participant	Measure (Total HRQoL)	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	Child	75	54.35	70.65	100	57.61
	Parent	89.13	57.61	79.35	81.52	60.87
2	Child	61.96	89.13	77.17	83.70	80.43
	Parent	77.17	67.39	95.65	86.96	82.61
3	Child	67.39	95.65	95.65	97.83	93.48
	Parent	39.13	95.65	93.48	93.48	93.48
4	Child	44.57	98.91	61.96	61.96	67.39
	Parent	71.74	95.65	81.52	100	76.09
5	Child	59.78	95.65	97.83	97.83	89.13
	Parent	79.35	50	93.48	97.83	95.65
6	Child	46.74	50	52.17	-	-
	Parent	59.78	56.52	67.39	-	-
7	Child	29.35	46.74	55.43	78.26	67.39
	Parent	33.70	50	53.26	61.96	66.30
8	Child	68.75	65.22	-	-	-
	Parent	78.26	86.96	-	-	-
Overall Means	Child	56.69 (15.24)	74.46 (22.58)	72.98 (18.33)	86.59 (14.95)	72.39 (12.38)
	Parent	66.03 (20.09)	69.97 (19.80)	80.59 (15.72)	86.96 (14.04)	76.30 (13.71)

Higher scores indicated better HRQoL

Table 8. Revised Children's Manifest Anxiety Scale Raw Scores

Participant	Measure	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	Total Anxiety	7	4	6	5	5
2	Total Anxiety	9	5	3	4	3
3	Total Anxiety	2	1	1	1	3
4	Total Anxiety	19*	19*	15	16	11
5	Total Anxiety	10	1	1	1	2
6	Total Anxiety	21*	18	23*	-	-
7	Total Anxiety	15	24*	12	23*	26*
8	Total Anxiety	12	11	-	-	-
Overall Means		12.27 (5.53)	10.38 (8.98)	8.71 (8.30)	8.33 (9.07)	8.33 (9.24)

*Clinically Significant (score of 19-28)

Table 9. Children's Depression Inventory T- Scores

Participant	Measure	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	Total CDI Symptoms	40	37	40	40	39
2	Total CDI Symptoms	42	38	36	44	39
3	Total CDI Symptoms	39	46	35	35	35
4	Total CDI Symptoms	56	44	42	53	41
	Negative Self-Esteem	70**	45	40	60*	45
5	Total CDI Symptoms	56	37	39	40	37
	Anhedonia	65*	38	38	42	38
6	Total CDI Symptoms	62*	50	62*	-	-
	Ineffectiveness	66*	59	59	-	-
7	Total CDI Symptoms	56	58	46	49	52
	Anhedonia	64*	67*	52	48	60*
8	Total CDI Symptoms	46	39	-	-	-
Overall Means		48.17 (8.64)	43.33 (8.14)	39.67 (4.03)	39.67 (4.03)	40.5 (5.99)

** Clinically Significant (T-Scores >70)
*At-Risk (T-Scores 60 – 70)

Table 10. Coping Strategies Questionnaire-Revised Factor Scores

Participant	Measure	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	**Coping Attempts**	51	64	49	28	63
	Negative Thinking	58	41	38	33	45
	Passive Adherence	88	74	70	62	73
2	**Coping Attempts**	94	112	110	121	124
	Negative Thinking	33	7	12	9	10
	Passive Adherence	91	92	90	90	99
3	**Coping Attempts**	60	89	95	77	22
	Negative Thinking	36	26	45	32	8
	Passive Adherence	57	81	92	85	45
4	**Coping Attempts**	39	78	85	127	106
	Negative Thinking	48	73	52	83	46
	Passive Adherence	63	76	67	92	72
5	**Coping Attempts**	74	54	78	71	58
	Negative Thinking	92	15	35	27	57
	Passive Adherence	87	78	84	111	106
6	**Coping Attempts**	84	68	61	-	-
	Negative Thinking	72	44	46	-	-
	Passive Adherence	85	68	64	-	-
7	**Coping Attempts**	76	158	142	41	107
	Negative Thinking	62	100	77	60	78
	Passive Adherence	94	132	120	117	110
8	**Coping Attempts**	48	89	-	-	-
	Negative Thinking	35	76	-	-	-
	Passive Adherence	78	103	-	-	-

Overall Means	**Coping Attempts**	65.75 (19.22)	89.00 (33.17)	88.57 (31.11)	77.50 (40.42)	91.60 (29.33)
	Negative Thinking	54.50 (20.73)	47.75 (32.58)	43.57 (19.57)	40.67 (26.42)	47.20 (24.67)
	Passive Adherence	80.13 (13.36)	88.00 (20.93)	77.50 (40.42)	92.83 (19.67)	92.00 (18.23)

Coping Attempts – higher scores suggest improvements in this positive coping strategy
Negative Thinking – lower scores suggest less of these negative coping strategies
Passive Adherence – higher scores suggest more adherence to medical recommendations

Table 11. Child Spiritual Coping Survey - Religious and Existential Frequency and Efficacy Scores

Participant	Measure	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	RC Frequency	29	11	17	18	6
	RC Efficacy	28	12	21	20	6
	EC Frequency	10	12	16	24	8
	EC Efficacy	10	12	14	18	4
2	RC Frequency	18	24	13	7	13
	RC Efficacy	21	18	8	7	12
	EC Frequency	36	32	25	17	33
	EC Efficacy	32	30	21	16	23
3	RC Frequency	0	28	30	23	22
	RC Efficacy	0	24	28	20	23
	EC Frequency	0	28	25	28	30
	EC Efficacy	0	23	22	25	30
4	RC Frequency	28	26	34	33	30
	RC Efficacy	20	16	-	31	34
	EC Frequency	29	35	52	34	25
	EC Efficacy	19	35	-	31	42
5	RC Frequency	36	36	34	36	32
	RC Efficacy	32	36	29	36	29
	EC Frequency	33	38	50	52	49
	EC Efficacy	34	38	48	52	41
6	RC Frequency	28	24	27	-	-
	RC Efficacy	36	25	26	-	-
	EC Frequency	29	25	27	-	-
	EC Efficacy	26	22	24	-	-
7	RC Frequency	33	36	30	26	31
	RC Efficacy	25	36	35	30	26
	EC Frequency	42	48	25	35	39
	EC Efficacy	28	48	26	28	40
8	RC Frequency	30	26	-	-	-
	RC Efficacy	30	35	-	-	-
	EC Frequency	27	30	-	-	-
	EC Efficacy	31	36	-	-	-

Overall Means	RC Frequency	25.25 (11.45)	26.38 (7.89)	26.43 (8.26)	23.83 (10.53)	22.40 (12.05)
	RC Efficacy	24.00 (11.1)	25.25 (9.57)	24.50 (9.27)	24.00 (10.49)	21.40 (11.87)
	EC Frequency	25.75 (13.92)	31.00 (10.43)	31.43 (13.84)	31.67 (11.98)	30.80 (15.47)
	EC Efficacy	22.50 (12.02)	30.50 (11.24)	25.83 (11.6)	28.33 (12.94)	30.00 (16.51)

Higher Scores indicate greater use (frequency) and perceived effectiveness (efficacy)

Table 12. Child Behavior Checklist T-Scores

Participant	Measure	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	Internalizing	58	57	61*	47	47
	Externalizing	56	43	48	34	34
	Total Problems	55	45	55	34	34
2	Internalizing	65**	68**	33	43	39
	Externalizing	55	51	40	44	44
	Total Problems	63*	65**	37	50	45
3	Internalizing	61*	52	48	-	
	Externalizing	50	40	44	-	
	Total Problems	53	41	41	-	
4	Internalizing	59	34	34	44	34
	Externalizing	40	34	34	34	34
	Total Problems	53	38	31	34	34
5	Internalizing	39	39	43	33	33
	Externalizing	47	47	34	34	34
	Total Problems	44	41	38	25	25
6	Internalizing	60*	60*	50	-	-
	Externalizing	59	59	51	-	-
	Total Problems	58	58	51	-	-
7	Internalizing	70**	67**	59	58	58
	Externalizing	57	51	51	51	50
	Total Problems	64**	59	53	51	51
8	Internalizing	39	39	-	-	-
	Externalizing	44	34	-	-	-
	Total Problems	41	42	-	-	-
Overall Means	Internalizing	56.38 (11.39)	52 (13.27)	46.86 (11.04)	45 (8.97)	42.2 (10.43)
	Externalizing	51 (6.85)	44.88 (8.81)	43.14 (7.36)	39.4 (7.8)	39.2 (7.43)
	Total Problems	53.88 (8.18)	48.63 (10.35)	43.17 (9.25)	38.8 (11.3)	37.8 (10.23)

**Clinically Significant (T-Score >64; >90th percentile)
*At-Risk (T-Score 60-63; 84th – 90th percentile)

Table 13. Adult Responses to Children's Symptoms Factor Scores

Participant	Measure	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	Protect	2	2.40	1.93	.80	1.53
	Minimize	1.33	.83	1.17	1	1.17
	Distract and Monitor	3.13	2.88	2.38	1.5	2.63
2	Protect	2	2.07	1.20	1.67	1.53
	Minimize	.50	.67	.50	.50	.50
	Distract and Monitor	3.25	2.25	1.63	2.25	2.25
3	Protect	3	1.93	1.80	2.67	2.53
	Minimize	1.67	1.17	.50	1	.67
	Distract and Monitor	3	2.25	3.25	2.75	2.75
4	Protect	3.67	3.27	4	3.87	3.87
	Minimize	.17	1.17	.67	0	1.33
	Distract and Monitor	3.63	3.75	4	4	4
5	Protect	2.67	3.07	3.87	4	3.87
	Minimize	1	1	0	.83	1
	Distract and Monitor	3.88	2.88	4	4	4
6	Protect	1	1.13	1.07	-	-
	Minimize	.33	.17	.17	-	-
	Distract and Monitor	2.13	1.38	1.75	-	-
7	Protect	3.33	2.40	2.50	2.60	2.27
	Minimize	.83	1	.67	.83	.17
	Distract and Monitor	3.75	3.38	2.63	3.13	3.13
8	Protect	3.80	2.87	-	-	-
	Minimize	1.67	0	-	-	-
	Distract and Monitor	4	3.13	-	-	-
Overall Means	Protect	2.68 (.96)	2.68 (.96)	2.34 (1.19)	2.6 (1.24)	2.6 (1.06)
	Minimize	.94 (.58)	.94 (.58)	.52 (.38)	.69 (.39)	.81 (.44)
	Distract and Monitor	3.34 (.61)	3.34 (.61)	2.80 (.98)	2.94 (.99)	3.13 (.73)

Higher scores indicate more of that kind of behavior

Table 14. Pediatric Inventory for Parents Total Scores

Participant	Measure	Baseline	Post-Treatment	2-Month Follow-Up	4-Month Follow-Up	6-Month Follow-Up
1	**Total Frequency**	45	136	73	91	78
	Communication	11	27	13	19	16
	Medical Care	8	27	10	18	15
	Emotional Dist.	16	52	24	36	27
	Role Function	10	30	26	18	2
	Total Difficulty	44	121	70	91	55
	Communication	10	26	13	21	15
	Medical Care	8	21	9	16	14
	Emotional Dist.	16	48	24	32	24
	Role Function	10	26	24	22	2
2	**Total Frequency**	49	71	49	46	42
	Communication	9	11	9	9	9
	Medical Care	8	9	8	8	8
	Emotional Dist.	20	29	16	17	15
	Role Function	12	22	16	1	10
	Total Difficulty	48	62	44	44	42
	Communication	9	9	9	9	9
	Medical Care	8	8	8	8	8
	Emotional Dist.	19	28	16	16	15
	Role Function	12	17	11	1	10
3	**Total Frequency**	112	55	91	45	43
	Communication	25	13	22	10	10
	Medical Care	21	10	16	9	8
	Emotional Dist.	39	21	31	16	15
	Role Function	27	11	22	10	10
	Total Difficulty	108	64	89	45	42
	Communication	18	12	20	9	9
	Medical Care	22	12	17	10	8
	Emotional Dist.	24	27	31	16	15
	Role Function	26	13	21	10	10
4 & 5	**Total Frequency**	86	72	88	102	60
	Communication	15	14	18	20	11
	Medical Care	12	13	19	25	9
	Emotional Dist.	38	26	29	31	26
	Role Function	21	19	22	4	14
	Total Difficulty	108	87	90	61	-

	Communication	17	16	19	9	-
	Medical Care	13	13	13	8	-
	Emotional Dist.	56	37	39	31	-
	Role Function	26	21	19	1	-
6	**Total Frequency**	65	84	79	-	-
	Communication	12	17	16	-	-
	Medical Care	11	15	13	-	-
	Emotional Dist.	28	34	33	-	-
	Role Function	14	18	17	-	-
	Total Difficulty	61	83	78	-	-
	Communication	11	17	14	-	-
	Medical Care	10	14	13	-	-
	Emotional Dist.	27	24	34	-	-
	Role Function	13	18	17	-	-
7	**Total Frequency**	126	124	101	111	100
	Communication	22	25	21	20	18
	Medical Care	26	23	18	18	17
	Emotional Dist.	55	53	40	50	44
	Role Function	23	23	22	23	21
	Total Difficulty	124	102	97	92	87
	Communication	22	15	12	13	13
	Medical Care	25	17	12	12	10
	Emotional Dist.	53	51	44	47	45
	Role Function	24	19	19	20	19
8	**Total Frequency**	64	71	-	-	-
	Communication	14	16	-	-	-
	Medical Care	10	12	-	-	-
	Emotional Dist.	22	25	-	-	-
	Role Function	18	18	-	-	
	Total Difficulty	63	99	-	-	-
	Communication	14	16	-	-	-
	Medical Care	10	17	-	-	-
	Emotional Dist.	22	37	-	-	-
	Role Function	17	29	-	-	-
Overall Means	Total Frequency	79.13 (28.97)	85.63 (28.66)	81.29 (16.78)	82.83 (29.61)	63.83 (22.13)
	Total Difficulty	84 (32.97)	88.13 (19.58)	79.71 (18.12)	65.67 (21.33)	56.5 (21.24)

Higher scores indicate greater frequency and difficulty

Table 15. Participant Hospitalizations

	Participants							
	1	2	3	4	5	6	7	8
Hospitalizations During 12-months Prior to Intervention	4	0	0	0	2	3	0	2
Hospitalizations During Intervention	2	0	0	0	0	0	0	0
Hospitalization During Post-Treatment Follow-Ups	3	0	0	0	1	1	0	1

Figure 1. Transactional stress and coping model of adjustment to chronic illness. From Thompson, Gustafson, George, and Spock (1994)

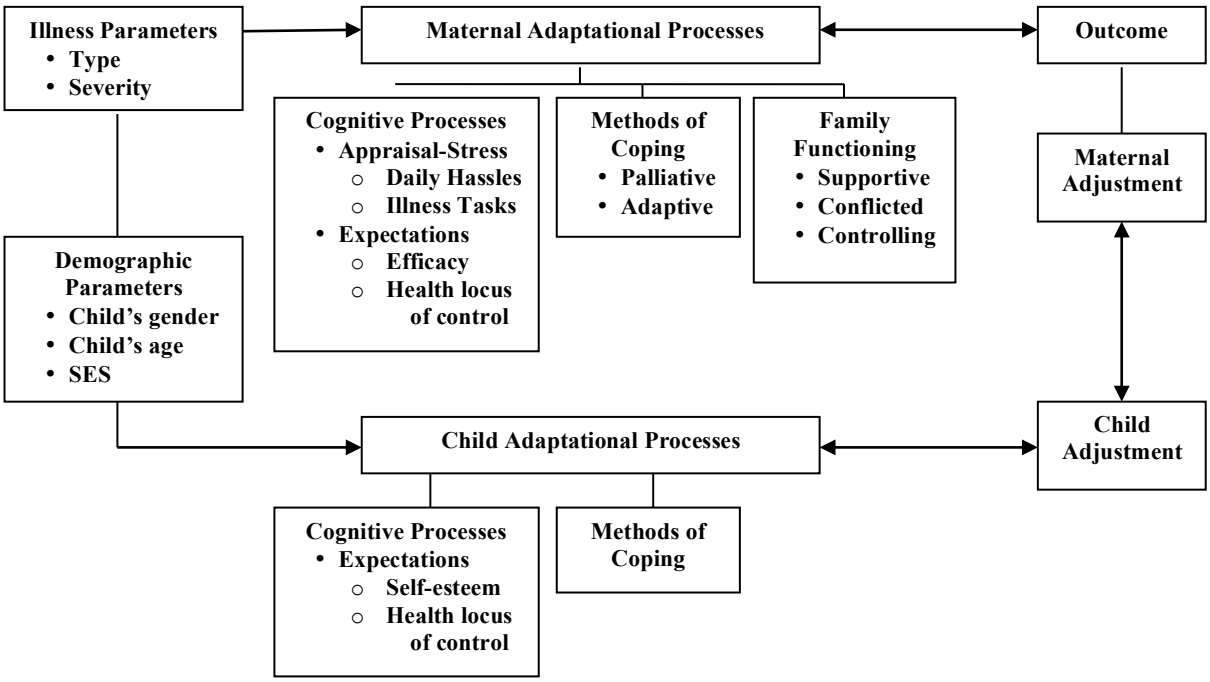


Figure 2. Participant Flow Chart

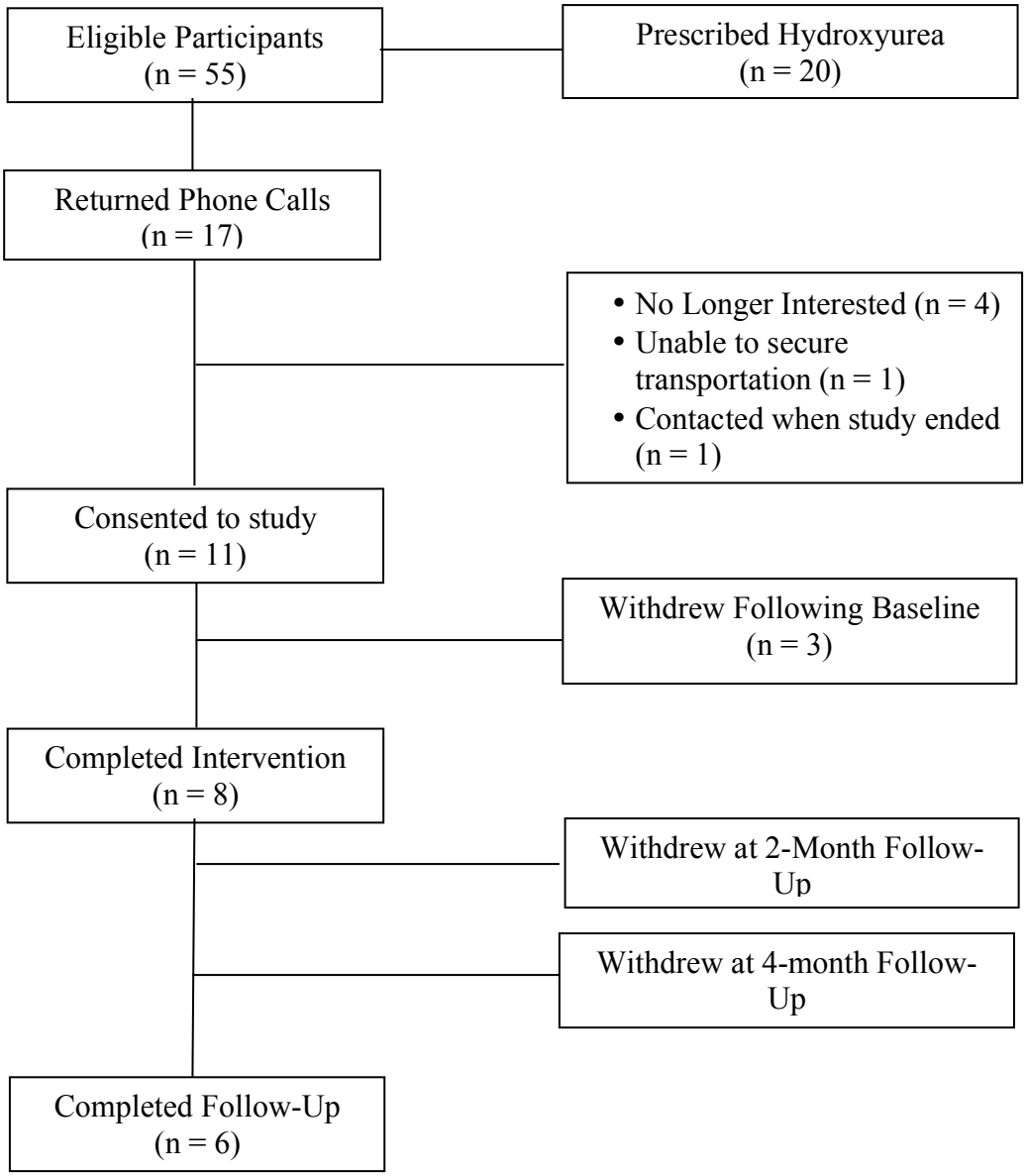
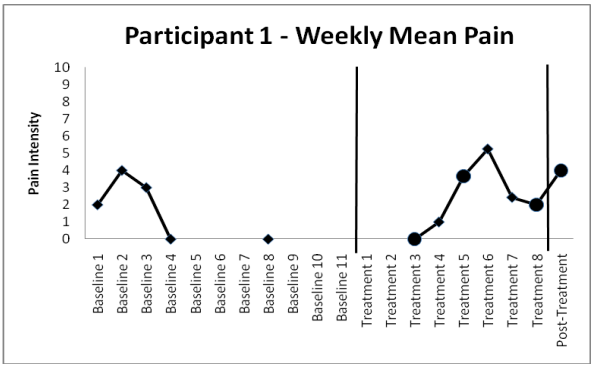
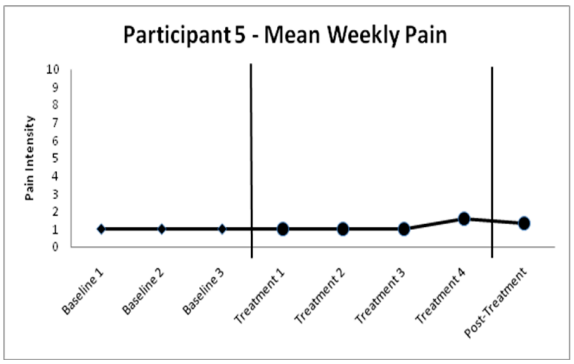
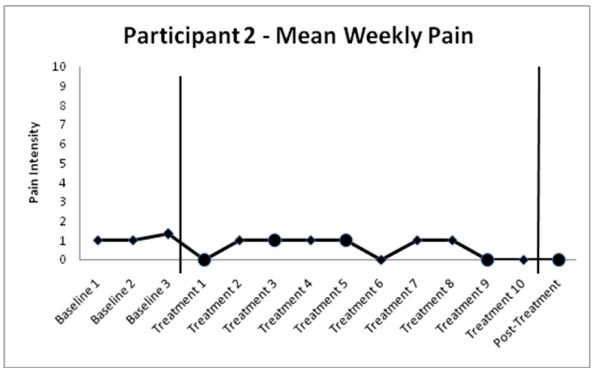
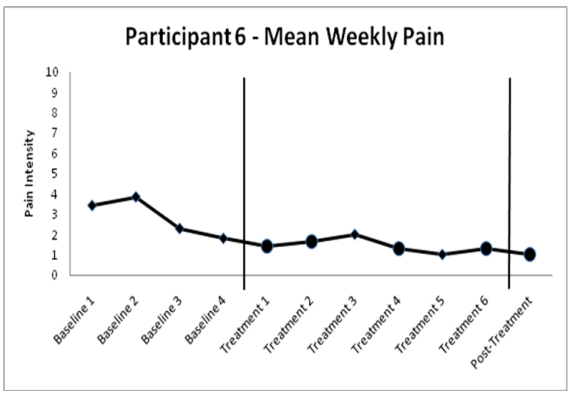


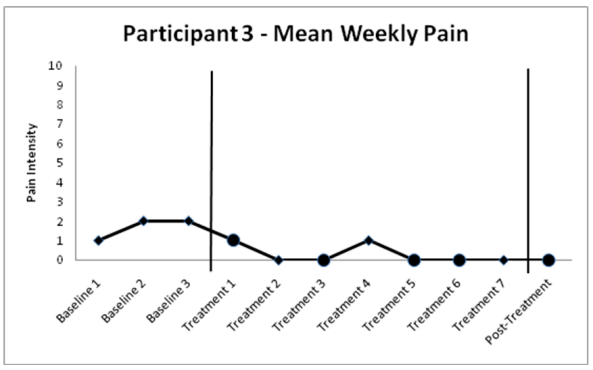
Figure 3. All Participants – Youth Weekly Mean Pain Intensity Ratings
(Circles represent Treatment Sessions)

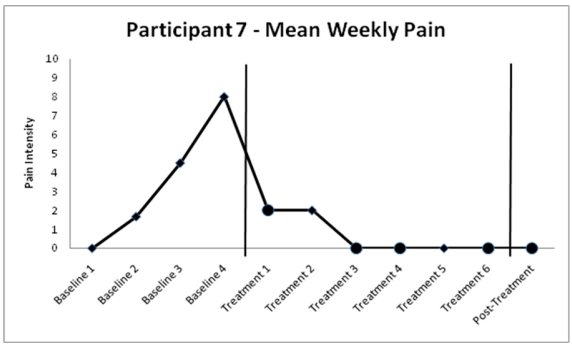


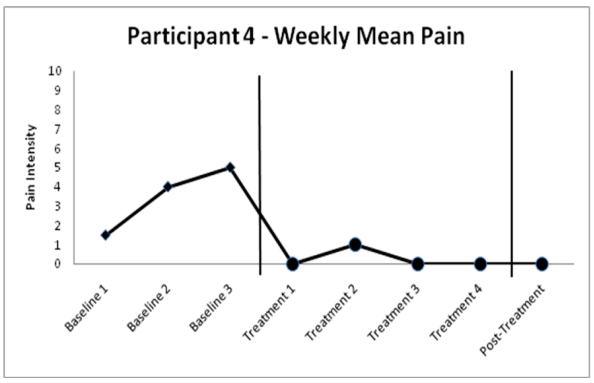












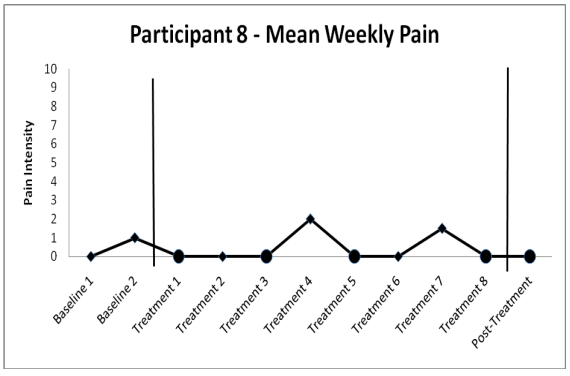
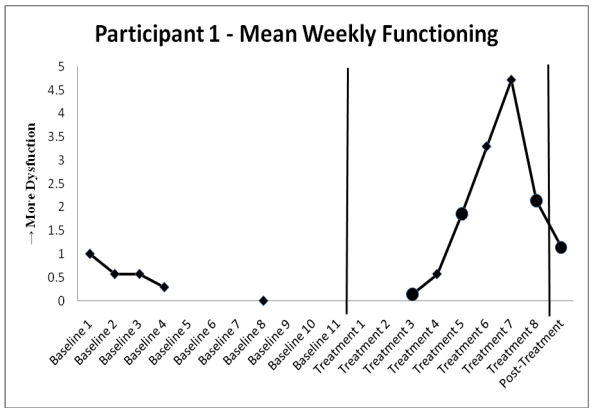
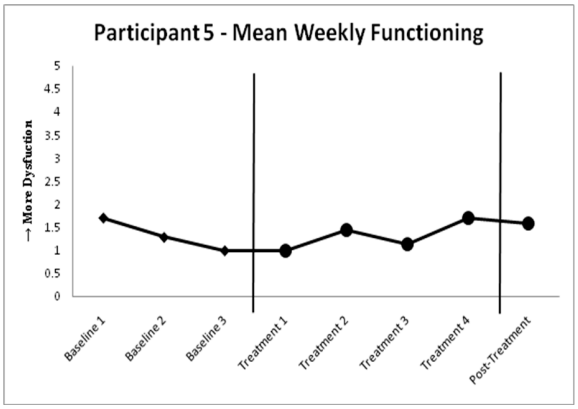
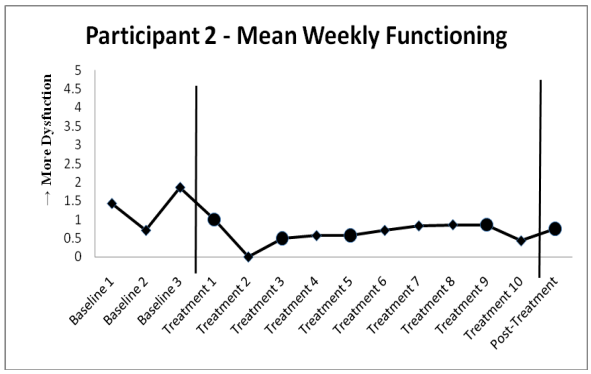
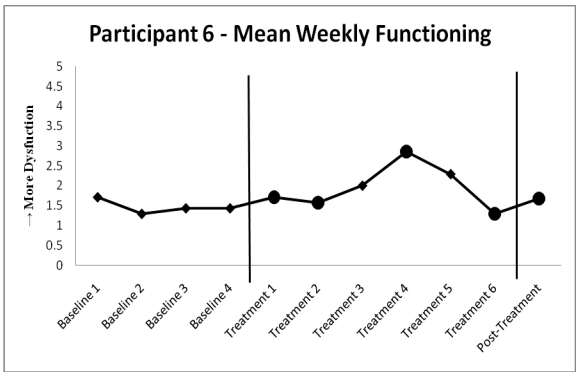


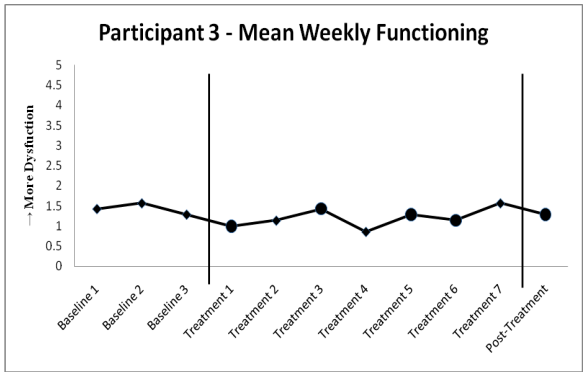
Figure 4. All Participants – Youth Weekly Mean Functioning
(Circles represent Treatment Sessions

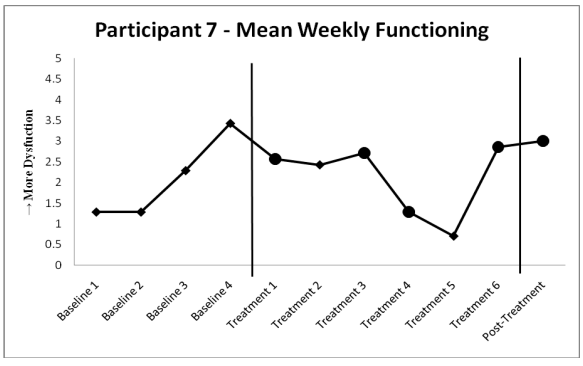


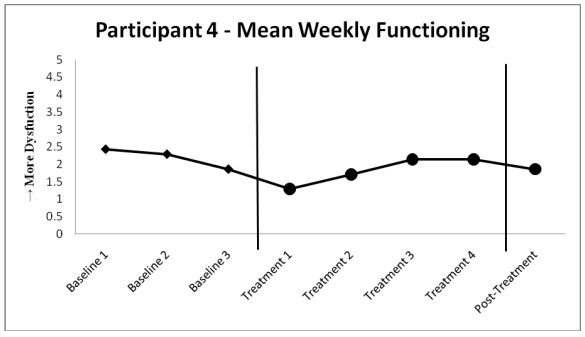












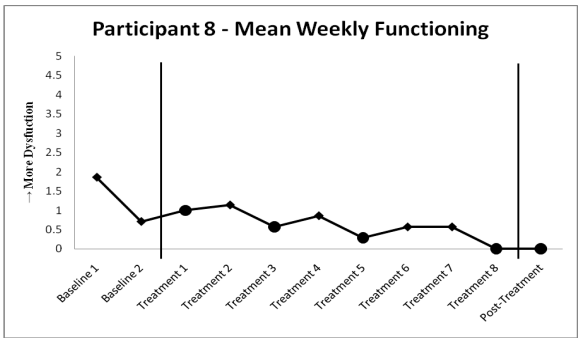
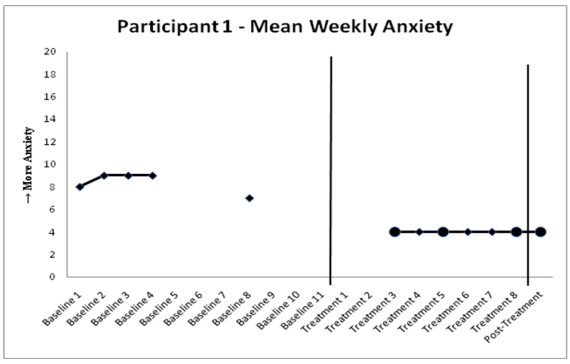
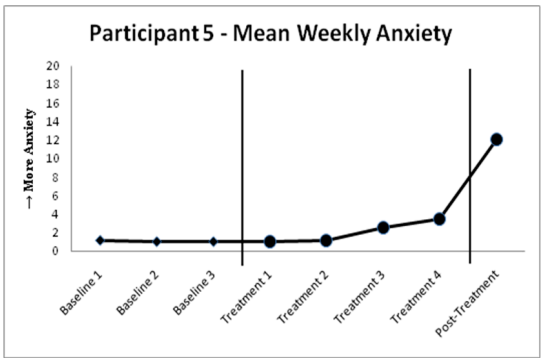
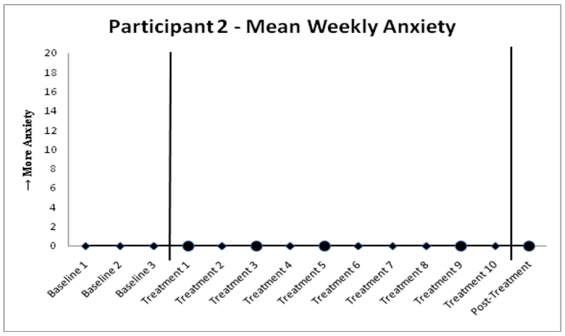
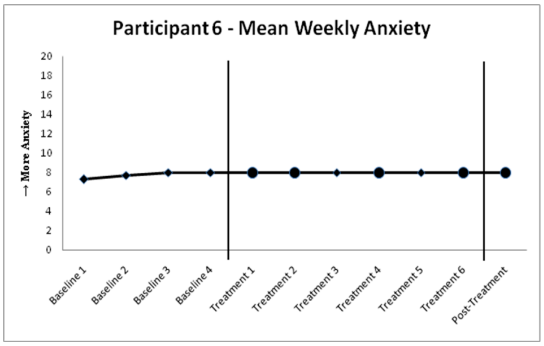


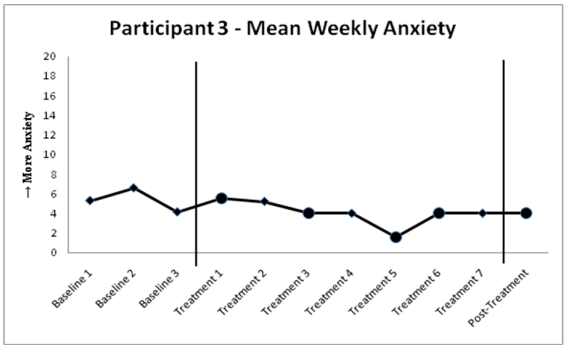
Figure 5. All Participants – Youth Weekly Mean Anxiety
(Circles represent Treatment Sessions

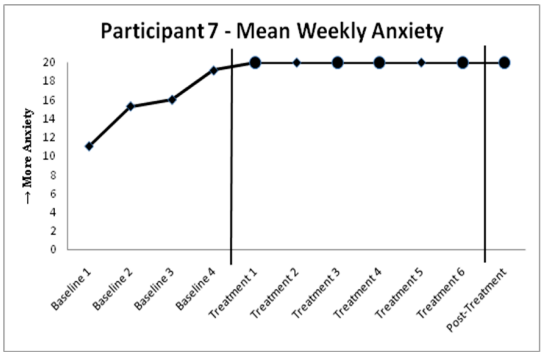


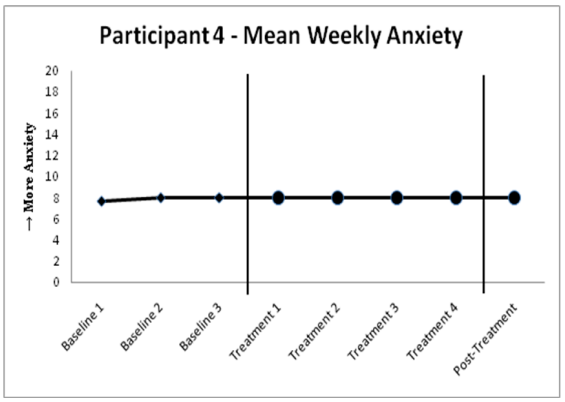


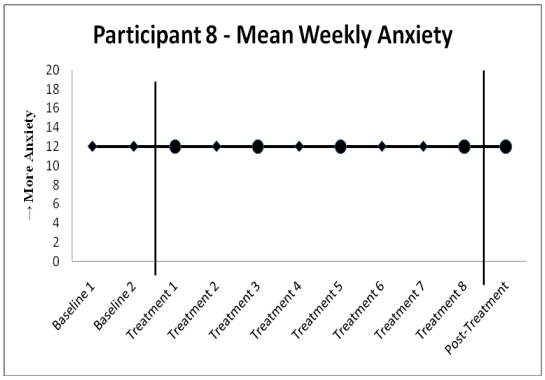












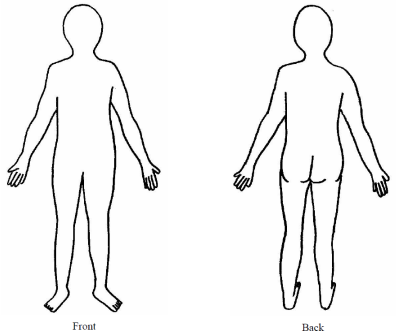
Appendix A

Daily Diary – Child Version

MONDAY (Date: __ / __ / ____)

ID# _____

IF YOU HURT, PUT AN X
WHERE IT HURT



1. What is your pain level now?

Not Hurting
No Discomfort
No Pain

Hurting a whole lot
Very Uncomfortable
Severe Pain

2. I worry when I am in pain.....	**0**	**1**	**2**	**3**	**4**	**5**
	Never					**Always**
3. I think that if my pain gets too bad it will never go away.....	**0**	**1**	**2**	**3**	**4**	**5**
	Never					**Always**
4. When I feel pain, I am afraid that something terrible will happen.....	**0**	**1**	**2**	**3**	**4**	**5**
	Never					**Always**
5. Pain is scary.....	**0**	**1**	**2**	**3**	**4**	**5**
	Never					**Always**

6. Did you go to school today?	**Yes**	**No**
7. Did you participate in any activities besides school today?	**Yes**	**No**
8. Did you miss any activities today?	**Yes**	**No**
What did you miss?		
9. Did you see friends today?	**Yes**	**No**
10. Did you do your chores today?	**Yes**	**No**
11. Did you see a doctor or nurse today?	**Yes**	**No**

12. Did you take your hydroxy- medicine today?	**Yes**	**No**
If yes, did someone remind you to take it?	**Yes**	**No**
If yes, who reminded you?		
13. Did you take any other medicine to help with your pain?	**Yes**	**No**

If yes, please check any medicine that you took: ❑ Penicillin ❑Folic acid ❑Tylenol ❑Ibuprofen

❑ Other: _____

14. Did you do anything else to help yourself with the pain? _____

Appendix B

WAKE FOREST UNIVERSITY HEALTH SCIENCES – DEPARTMENT OF HEMATOLOGY

Parental Permission for Participants in Research Projects Involving Human Subject – Parent

Title: A Family-Based Cognitive-Behavioral Intervention for Pediatric Patients with Sickle Cell Disease
Principal Investigators: Alexandra Boeving Allen, Ph.D.
Study Coordinator: Rachel Moore, M.S.

I. Purpose of this Research/Project
We want to know what kids and parents think and feel about the pain and other signs of Sickle Cell Disease (SCD), so we are doing a research study which you and your child are invited to take part in. Our goal is to study your child's thoughts, behaviors, and feelings about SCD pain. We also want to study other related effects, (like adjustment problems, activity levels, coping skills.) We are studying these things in the hope of making your child's quality of life and health outcomes better. We will give your child information about SCD and we will teach your child things to do that will help your child deal with his/her pain. We will teach your child how to relax, how to keep track of his/her feelings, and how to set goals. We will also talk to you about how you and your family support your child. We hope that your help will let us help other children and families deal with SCD.

This "Informed Consent Form" gives you details about the study to help you decide if you wish to be in the study. This form also tells you about your rights and the rights of your child if you choose to be in this study. It is important to us that you know this information. Please ask questions if you would like us to explain any of the items in this form. You are free to talk with friends and family as you make your mind up about being in the study. You will be given a signed copy of this consent form.

II. Procedures
During the study, we will ask you and your child to fill out some forms. We will ask you to rate your child's pain, activity levels, health care use, quality of life, emotions, and family roles. We will also ask you for information about you and your family such as age, education, ethnicity, and income. We can help you with reading and filling out the forms.

Before the study starts, we will ask you and your child to fill out daily pain records for 2 to 5 weeks to learn more about the problems your child may have related to his/her SCD. After filling out these daily records, you will begin the study and we will ask you to come for 5 visits, scheduled twice a month for about 10 weeks. We will ask you and your child to come to all of these visits. During the study we will ask you and your child to complete the daily pain records, as well as brief homework.

After these visits, we will give you and your child the same forms to fill out. You can fill out these forms during your child's monthly checkup. It should take about 60 minutes (1 hour) to fill out these forms. Four to 6 months after these visits, we will send you and your child the same forms to fill out. We will provide an envelope with stamps so that you can return the forms by mail.

As part of the study, we will look at your child's health records to get basic information related to his/her illness and the number of doctor visits in the past year.

As part of this study, we will audiotape all of our visits with you and your child. The tapes will help us make sure that we are doing a good job with the study. You can ask that the tape be stopped at any time

during the study. But, you will not be able to check or look at the audio-tapes before they are used in this study.

Please choose one of the choices about the use of audio-tapes in this research study:
_____ I would like the audiotapes of me and my child to be destroyed once their use in this study is done.

_____ The audiotapes of me can be kept for use in future studies. The tapes will be kept secure and future studies will be reviewed by an IRB. I understand I will not be able to look at or approve their future use.

III. Risks
The risks of being in this study are minor; being in this study is about as risky as usual mental and emotional evaluation. You and your child will be asked to talk about thoughts and feelings related to his/her disease. It may be hard for you or your child to talk about these things, but the study will happen in a caring setting. We are trained in helping people in crisis and a family member will be present. If you or your child get upset or cannot continue, we will give you support and help. If you or your child seems to be a risk to themselves or others, we will also help you.

IV. Benefits
Being in this study may improve you and your child's quality of life. From being in the study, you and your child may learn things to do to deal with pain and the problems your child has because of pain. Your child may also see decreases in pain, and may be more active because of being in the study. Thus, the gains of the study seem to be much greater than the possible risks.

V. Confidentiality
We will not tell or share information with anyone else. We will never give your information to anyone without getting written permission from you first. However, the results of this study may be used for scientific and/or learning purposes, talked about at scientific meetings, and/or are made available in print. Whenever we use or discuss the study results, we will take out all of your private information. Audiotapes of all of the visits will also not be shared with anyone else. We will give tapes a number, and the tapes will be kept in a locked area with the rest of the study data. You and your child's name will not be on the tapes. The tapes and other data will be kept for at least 5 years after the end of the study, at which time they will be destroyed. We will not give these tapes or any other information we from you to anyone who is not part of the study unless we get written consent from you or your child first. The only time we would tell someone about what you or your child said was if you or your child let us know that one of you were a danger to yourself, someone else, or if someone else was hurting you. We hope you will talk to your child about his/her experience in the study.

VI. Payment
There are no costs for being in this study. Any medical costs that are not related to this study, like your child's regular medical care, are not covered by the study. We will give your family $10 for the first visit. We will also give your family $10 for each visit that you and your child take part in. We will also give your family $20 for filling out the forms at 2-, 4-, and 6-months after the study visits end.

VII. Freedom to Withdraw
Being in this study is voluntary. You may choose not to take part in the study or you may also choose to leave the study at any time. You will not get any fine or loss of benefits if you decide not to be in the study or if you decide to leave the study. You hold these rights when you decide to be in the study. You may choose to not answer any question you do not wish to answer. If you decide to stop this study we hope you talk with us first.

VIII. Participant's Responsibilities

I willingly agree to be in this study and to let my child be in this study. I have the following duties: 1. Let the researchers know if I am uncomfortable or wish to stop the study at any time; 2. Answer honestly any questions I choose to answer; 3. Attend all visits with my child as long as I am willing to participate; 4. Discuss the study with my child, if I wish, during and after the study.

IX. Participant's Permission

I have read and understand the Informed Consent and details of this study. I have had a chance to ask questions, and my questions have been answered. I understand that I have the right to end this study for any reason if I so choose without penalty. If I have not already been given a copy of the Privacy Notice, I may ask for one or one will be given to me. This study has been approved by Wake Forest University. If I have any questions regarding this study, I should contact one of the people named below:

Dr. Alexandra Boeving Allen, Principal Investigator 336-716-1284
Rachel Moore, M. S., Study Coordinator 540-231-8504
Wake Forest IRB Chairman 336-716-4542

I hereby acknowledge the above and give my voluntary permission for the participation of my child _____ (write in child's name) in this study. By signing this consent and authorization form, I am not releasing or agreeing to release the researchers, the institution or its agents from liability for negligence.

Signature of Parent/Guardian: _____ Date: _____

Printed Name of Parent/Guardian: _____

Person Obtaining Consent: _____ Date: _____

Printed Name of Person Obtaining Consent: _____

WAKE FOREST UNIVERSITY HEALTH SCIENCES – DEPARTMENT OF HEMATOLOGY

Informed Consent for Participants in Research Projects Involving Human Subject

Title: A Family-Based Cognitive-Behavioral Intervention for Pediatric Patients with Sickle Cell Disease
Principal Investigators: Alexandra Boeving Allen, Ph.D.
Study Coordinator: Rachel Moore, M.S.

I. Purpose of this Research/Project
We want to know what kids and parents think and feel about the pain and other signs of Sickle Cell Disease (SCD), so we are doing a research study which you and your child are invited to take part in. Our goal is to study your child's thoughts, behaviors, and feelings about SCD pain. We also want to study other related effects, (like adjustment problems, activity levels, coping skills.) We are studying these things in the hope of making your child's quality of life and health outcomes better. We will give your child information about SCD and we will teach your child things to do that will help your child deal with his/her pain. We will teach your child how to relax, how to keep track of his/her feelings, and how to set goals. We will also talk to you about how you and your family support your child. We hope that your help will let us help other children and families deal with SCD.

This "Informed Consent Form" gives you details about the study to help you decide if you wish to be in the study. This form also tells you about your rights and the rights of your child if you choose to be in this study. It is important to us that you know this information. Please ask questions if you would like us to explain any of the items in this form. You are free to talk with friends and family as you make your mind up about being in the study. You will be given a signed copy of this consent form.

II. Procedures
During the study, we will ask you and your child to fill out some forms. We will ask you to rate your child's pain, activity levels, health care use, quality of life, emotions, and family roles. We will also ask you for information about you and your family such as age, education, ethnicity, and income. We can help you with reading and filling out the forms.

Before the study starts, we will ask you and your child to fill out daily pain records for 2 to 5 weeks to learn more about the problems your child may have related to his/her SCD. After filling out these daily records, you will begin the study and we will ask you to come for 5 visits, scheduled twice a month for about 10 weeks. We will ask you and your child to come to all of these visits. During the study we will ask you and your child to complete the daily pain records, as well as brief homework.

After these visits, we will give you and your child the same forms to fill out. You can fill out these forms during your child's monthly checkup. It should take about 60 minutes (1 hour) to fill out these forms. Four to 6 months after these visits, we will send you and your child the same forms to fill out. We will provide an envelope with stamps so that you can return the forms by mail.

As part of the study, we will look at your child's health records to get basic information related to his/her illness and the number of doctor visits in the past year.

As part of this study, we will audiotape all of our visits with you and your child. The tapes will help us make sure that we are doing a good job with the study. You can ask that the tape be stopped at any time during the study. But, you will not be able to check or look at the audio-tapes before they are used in this study.

Please choose one of the choices about the use of audio-tapes in this research study:

_____ I would like the audiotapes of me and my child to be destroyed once their use in this study is done.

_____ The audiotapes of me can be kept for use in future studies. The tapes will be kept secure and future studies will be reviewed by an IRB. I understand I will not be able to look at or approve their future use.

III. Risks
The risks of being in this study are minor; being in this study is about as risky as usual mental and emotional evaluation. You and your child will be asked to talk about thoughts and feelings related to his/her disease. It may be hard for you or your child to talk about these things, but the study will happen in a caring setting. We are trained in helping people in crisis and a family member will be present. If you or your child get upset or cannot continue, we will give you support and help. If you or your child seems to be a risk to themselves or others, we will also help you.

IV. Benefits
Being in this study may improve you and your child's quality of life. From being in the study, you and your child may learn things to do to deal with pain and the problems your child has because of pain. Your child may also see decreases in pain, and may be more active because of being in the study. Thus, the gains of the study seem to be much greater than the possible risks.

V. Confidentiality
We will not tell or share information with anyone else. We will never give your information to anyone without getting written permission from you first. However, the results of this study may be used for scientific and/or learning purposes, talked about at scientific meetings, and/or are made available in print. Whenever we use or discuss the study results, we will take out all of your private information. Audiotapes of all of the visits will also not be shared with anyone else. We will give tapes a number, and the tapes will be kept in a locked area with the rest of the study data. You and your child's name will not be on the tapes. The tapes and other data will be kept for at least 5 years after the end of the study, at which time they will be destroyed. We will not give these tapes or any other information we from you to anyone who is not part of the study unless we get written consent from you or your child first. The only time we would tell someone about what you or your child said was if you or your child let us know that one of you were a danger to yourself, someone else, or if someone else was hurting you. We hope you will talk to your child about his/her experience in the study.

VI. Payment
There are no costs for being in this study. Any medical costs that are not related to this study, like your child's regular medical care, are not covered by the study. We will give your family $10 for the first visit. We will also give your family $10 for each visit that you and your child take part in. We will also give your family $20 for filling out the forms at 2-, 4-, and 6-months after the study visits end.

VII. Freedom to Withdraw
Being in this study is voluntary. You may choose not to take part in the study or you may also choose to leave the study at any time. You will not get any fine or loss of benefits if you decide not to be in the study or if you decide to leave the study. You hold these rights when you decide to be in the study. You may choose to not answer any question you do not wish to answer. If you decide to stop this study we hope you talk with us first.

VIII. Participant's Responsibilities

I willingly agree to be in this study. I have the following duties: 1. Let the researchers know if I am uncomfortable or wish to stop the study at any time; 2. Answer honestly any questions I choose to answer; 3. Attend all visits with my child as long as I am willing to participate; 4. Discuss the study with my child, if I wish, during and after the study.

IX. Participant's Permission

I have read and understand the Informed Consent and details of this study. I have had a chance to ask questions, and my questions have been answered. I understand that I have the right to end this study for any reason if I so choose without penalty. If I have not already been given a copy of the Privacy Notice, I may ask for one or one will be given to me. This study has been approved by Wake Forest University. If I have any questions regarding this study, I should contact one of the people named below:

Dr. Alexandra Boeving Allen, Principal Investigator 336-716-1284
Rachel Moore, M. S., Study Coordinator 540-231-8504
Wake Forest IRB Chairman 336-716-4542

I hereby acknowledge the above and give my voluntary consent for my participation in this study. By signing this consent and authorization form, I am not releasing or agreeing to release the investigator, the institution or its agents from liability for negligence.

Signature of Participant: _____ Date: _____

Printed Name of Participant: _____

Person Obtaining Consent: _____ Date: _____

Printed Name of Person Obtaining Consent: _____

WAKE FOREST UNIVERSITY HEALTH SCIENCES – DEPARTMENT OF HEMATOLOGY

Informed Consent for Participants in Research Projects Involving Human Subjects –Adolescents

Title: A Family-Based Cognitive-Behavioral Intervention for Pediatric Patients with Sickle Cell Disease
Principal Investigators: Alexandra Boeving Allen, Ph.D.
Study Coordinator: Rachel Moore, M.S.

I. Purpose of this Research/Project
We want to know what kids and parents think and feel about the pain and other signs of Sickle Cell Disease (SCD), so we are doing a research study. The paper I'm reading to you now is called a "Child Assent Form." This form tells about our study, and tells you what it will be like for you if you want to help us with our study. Your parent (or an adult in your family) says that it is OK for you to talk to us, but you do not have to be in our study if you do not want to. Helping with this study is your choice, and you may say no if you do not want to talk to us. You may ask questions any time you want.

II. Procedures
If you want to be in our study, you and your parent (or a grown-up in your family) will fill out some paper forms for us. We will ask you some questions about your pain, activities you do, what you are feeling, and how you deal with your pain. If you want, we can help you with reading and filling out these forms.

After you fill-out the paper forms, you and your parent (or a grown-up in your family) will be asked to fill out daily pain records for about 2 to 5 weeks. This will help us to get to know you better. Then, you and your parent (or a grown-up in your family) will begin the therapy. We will meet 5 times, twice a month for about 10 weeks. Both you and your parent (or a grown-up in your family) will be asked to come for all of these visits. At the visits, we will work together on ways to deal with your pain and other problems you may be having. We will also ask you and your parent (or a grown-up in your family) to fill out daily pain records, as well as do some brief homework.

All of the visits will be audio-recorded to make sure that we are doing the things that we need to during the sessions. At 2, 4, and 6 months after these visits, you and your parent (or a grown-up in your family) will be given the same packet of paper forms to fill out. As part of the research study, we would also like to look at your health records to find out the number of doctor visits you or your parent (or a grown-up in your family) made in the past year.

III. Risks
Being in this study is not dangerous or bad for you. If you feel bad while you are talking to us, you should tell us and we will help you.

IV. Benefits
By answering questions and joining our study, you will be helping us to better know lots of important things about kids with SCD. Also, by helping us out, you may be better able to deal with your SCD pain.

V. Confidentiality
This is a big word that means that anything you tell us we can't tell anyone else. We will take your answers to our questions, and we will add your answers together with the answers of other kids in our study. We will do a lot of math with the answers. We will never tell anyone what your own answers were, and no one will know that you were one of the kids in our study. The only time we would tell someone about what you said was if you let us know that you wanted to hurt yourself, someone else, or if someone

else was hurting you. During our study, and when you are done working with us, you should talk to your parent (or a grown-up in your family) about what it was like to be in a research study.

VI. Payment
All the families in our study will get $10 for the visit today. Also, for each visit that you and your parent (or a grown-up in your family) come to, your family will get $10. Families will also get money after filling out the group of paper forms at 2, 4, and 6 months after the end of the study. For each time you and your parent (or a grown-up in your family) fill this packet out you can earn $20.

VII. Freedom to Withdraw
You can stop being in our study any time, even in the middle. You do not have to answer questions you do not want to answer. Nothing bad will happen to you if you stop talking to us.

VIII. Approval of Research
There is a team of people at our medical center who make sure this study is safe for kids and will not hurt them. This team of people says that we can do this study and to ask you these questions.

IX. Participant's Permission
By writing your name below, you are saying that this paper has been read to you and that you get what we said in this paper. You know that you can ask questions anytime and can stop being in the study anytime. And, if you have more questions later, you can call – or you can have your parent (or a grown-up in your family) call – any of these people:

Dr. Alexandra Boeving Allen, Principal Investigator 336-716-1284
Rachel Moore, M. S., Study Coordinator 540-231-8504
Wake Forest IRB Chair 336-716-4542

Child's Name (written in by Child): _____

Child's Printed Name (by an Adult): _____

X. Parent Permission
By signing below, I indicate that my child has been read this form in front of me, and that my child's questions have been answered. My child has my permission to take part in this study. I believe that my child wants to take part in this study and that s/he knows s/he can stop whenever s/he wishes.

_____
Legally Authorized Representative Name (Print)

The above named Legally Authorized Representative has legal authority to act for the research subject based upon (specify health care power of attorney, spouse, parent, etc.)

_____
Relationship to the Subject

_____ _____
Legally Authorized Representative Signature Date

Signature of Person Obtaining Consent: _____ Date: _____
Printed Name of Person Obtaining Consent: _____

WAKE FOREST UNIVERSITY HEALTH SCIENCES – DEPARTMENT OF HEMATOLOGY

Informed Consent for Participants in Research Projects Involving Human Subjects – Child

Title: A Family-Based Cognitive-Behavioral Intervention for Pediatric Patients with Sickle Cell Disease
Principal Investigators: Alexandra Boeving Allen, Ph.D.
Study Coordinator: Rachel Moore, M.S.

I. Purpose of this Research/Project
We want to know what kids and families think and feel about Sickle Cell Disease (SCD), so we are doing a study. The paper I'm reading to you now tells you about our study, and tells you what it will be like for you if you want to help us. Your family says that it is OK for you to talk to us, but you do not have to be in our study if you do not want to. Nobody will be upset or mad if you don't want to be in the study. You may say no if you do not want to talk to us. You may ask questions any time you want.

II. Procedures
If you want to be in our study, your family will fill out some paper forms. We will ask you some questions about your pain, things you do, what you are feeling, and how you deal with your pain. If you want, we can help you with reading and writing.

You and your family will be asked to fill out daily pain sheets and do some brief homework. This will help us to get to know you better. Then, you and your family will begin therapy. You and your family will meet with me 5 times. At the visits, we will work together on ways to deal with your pain and other problems you may be having. All of the visits will be audio-recorded to make sure that we are doing the things that we need to do during the visits. After your visits, you and your family will be given the same paper forms to fill out again.

As part of the study, we would also like to look at your health records to find out how many times you visited your doctor.

III. Risks
Being in this study is not bad for you. If you feel bad while you are talking to us, we want you to tell us and we will help you.

IV. Benefits
By talking to us and filling out the paper forms, you will help us know lots of things about kids with SCD, and it may help you deal with your SCD pain.

V. Confidentiality
This is a big word that means that anything you tell us we can't tell anyone else. We will never tell anyone what your own answers are, and no one will know that you were one of the kids in our study. The only time we would tell someone about what you said was if you let us know that you wanted to hurt yourself, someone else, or if someone else was hurting you.

VI. Payment
All the families in our study will get $10 for the visit today. Also, for each visit that you and your family come to, your family will get $10. Your family will also get $20 for filling out the paper forms after you have had 6 visits.

VII. Freedom to Withdraw
You can stop being in our study any time, even in the middle. You do not have to answer questions you do not want to answer. Nothing bad will happen to you if you stop talking to us.

VIII. Approval of Research
There is a team of people at our health center who make sure this study is safe for kids and will not hurt them. This team of people says that we can do this study and ask you questions.

IX. Participant's Permission
By writing your name below, you are saying that this paper has been read to you and that you get what we said. You know that you can ask questions or stop the study at anytime. If you have more questions later, you can call – or you can have your family call – any of these people:

Dr. Alexandra Boeving Allen, Principal Investigator 336-716-1284
Rachel Moore, M. S., Study Coordinator 540-231-8504
Wake Forest IRB Chair 336-716-4542

Child's Name (written in by Child): _____

Child's Printed Name (by an Adult): _____

X. Parent Permission
By signing below, I indicate that my child has been read this form in front of me, and that my child's questions have been answered. My child has my permission to take part in this study. I believe that my child wants to take part in this study and that s/he knows s/he can stop at any time s/he wishes.

_____
Legally Authorized Representative Name (Print)

The above named Legally Authorized Representative has legal authority to act for the research subject based upon (specify health care power of attorney, spouse, parent, etc.)

_____
Relationship to the Subject

_____ _____
Legally Authorized Representative Signature Date

Signature of Person Obtaining Consent: _____ Date: _____

Printed Name of Person Obtaining Consent: _____

Virginia Tech Institutional Review Board: Project No. 09-523
Approved June 23, 2009 to June 22, 2010

WAKE FOREST
UNIVERSITY
HEALTH SCIENCES

Department/Section of Pediatric Hematology/Oncology

Assent Form to Participate in a Research Study

A FAMILY-BASED COGNITIVE-BEHAVIORAL INTERVENTION FOR PEDIATRIC PATIENTS WITH SICKLE CELL DISEASE

Alexandra Boeving Allen, Ph.D., Principal Investigator

Why am I here?

We want to tell you about a research study about children with Sickle Cell Disease. We want to see if you would like to be in this research study. Dr. Boeving Allen and some other people at this medical center are doing this study.

What will happen to me?

Only if you want to be in the study, the following things will happen:

- We want to know what kids and families think and feel about Sickle Cell Disease (SCD), so we are doing a study. The paper I'm reading to you now tells you about our study, and tells you what it will be like for you if you want to help us. Your family says that it is OK for you to talk to us, but you do not have to be in our study if you do not want to. Nobody will be upset or mad if you don't want to be in the study. You may say no if you do not want to talk to us. You may ask questions any time you want.

Will the study hurt?

Being in this study is not dangerous or bad for you. If you feel bad while you are talking to us, you should tell us and we will help you.

Will I get better if I am in the study?

By talking to us and filling out the paper forms, you will help us know lots of things about kids with SCD, and it may help you deal with your SCD pain.

What if I have questions?

You can ask questions any time. You can ask questions now, or later. You can talk to the doctors or others helping with the study. You can also talk with your parents or other adults about being in the study if you want to.

Do I have to be in the study?

You do not have to be in the study. No one will be mad at you or unhappy if I don't want to do this. If you don't want to be in this study, you just have to tell the study doctor. And if you want to be in the study, just let the study doctor know. You can say yes now and change your mind later. It's up to you.

_____	_____	_____
Signature of Subject	Age	Date
_____		_____
Person Obtaining Assent		Date

WFU Health Sciences
Institutional Review Board
IRB Number IRB00010023
Meeting Date Approved 9/25/2009
Version Valid Until 9/25/2010

WAKE FOREST
UNIVERSITY
HEALTH SCIENCES

Department/Section of Pediatric Hematology/Oncology

A Family-Based Cognitive-Behavioral Intervention for Pediatric Patients with Sickle Cell Disease
Informed Consent Form to Participate in Research
Alexandra Boeving Allen, Ph.D., Principal Investigator

Introduction
You and your child are invited to be in a research study. Research studies are designed to gain scientific knowledge that may help other people in the future. You are being asked to take part in this study because your child has Sickle Cell Disease. You and your child's participation is voluntary. Please take your time in making your decision as to whether or not you wish to participate. Ask the study staff to explain any words or information contained in this informed consent document that you do not understand. You and your child may also discuss the study with your friends and family.

Why Is This Study Being Done?
The purpose of this research study is to let us know what kids and parents think and feel about the pain and other signs of Sickle Cell Disease (SCD), so we are doing a research study which you and your child are invited to take part in. Our goal is to study your child's thoughts, behaviors, and feelings about SCD pain. We also want to study other related effects, (like adjustment problems, activity levels, coping skills.) We are studying these things in the hope of making your child's quality of life and health outcomes better. We will give your child information about SCD and we will teach your child things to do that will help your child deal with his/her pain. We will teach your child how to relax, how to keep track of his/her feelings, and how to set goals. We will also talk to you about how you and your family support your child. We hope that your help will let us help other children and families deal with SCD.

How Many People Will Take Part in the Study?
Nine children and adolescents at this research sites will take part in this study.

What Is Involved in the Study?
During the study, we will ask you and your to fill out some forms. We will ask you to rate your child's pain, activity levels, health care use, quality of life, emotions, and family roles. We will also ask you for information about you and your family such as age, education, ethnicity, and income. We can help you with reading and filling out the forms.

Before the study starts, we will ask you and your child to fill out daily pain records for 2 to 5 weeks to learn more about the problems your child may have related to his/her SCD. After filling out these daily records, you will begin the study and we will ask you to come for 5 visits, scheduled twice a month for about 10 weeks. We will ask you and your child to come to all of these visits. During the study we will ask you and your child to complete the daily pain records, as well as brief homework.

After these visits, we will give you and your child the same forms to fill out. You can fill out these forms during your child's monthly checkup. It should take about 60 minutes (1 hour) to fill out these forms.

Four to 6 months after these visits, we will send you and your child the same forms to fill out. We will provide an envelope with stamps so that you can return the forms by mail.

As part of the study, we will look at your child's health records to get basic information related to his/her illness and the number of doctor visits in the past year.

As part of this research study, you and your child will be audiotaped. This is being done to help us make sure that we are doing a good job with the study. You understand that you or your child may request the filming or recording be stopped at any time during the course of the research study. You or your child can also withdraw your consent to use and disclose the audiotape before it is used. You should also understand that you and your child will not be able to inspect, review, or approve the audiotapes or other media (including articles containing such) before they are used in this study.

Please choose one of the following regarding the use and disclosure of the audiotape used in this research study:

_____ I would like the audiotapes of me and my child to be destroyed once their use in this study is finished.

____ The audiotapes of me and my child can be kept for use in future studies provide they are kept secure and any future study will be reviewed by an IRB. I understand that I and my child will not be able to inspect, review or approve their future use.

How Long Will I Be in the Study?
You and your child will be in the active participation of this study for about 4 months. In addition, the study will involve long-term follow-ups (2-, 4-, and 6-months post-treatment).

You and your child can stop participating at any time. If you or your child decides to stop participating in the study we encourage you and your child to talk to the investigators or study staff first to learn about any potential health or safety consequences.

What Are the Risks of the Study?
The risks of being in this study are minor; being in this study is about as risky as usual mental and emotional evaluation. You and your child will be asked to talk about thoughts and feelings related to his/her disease. It may be hard for you or your child to talk about these things, but the study will happen in a caring setting. We are trained in helping people in crisis and a family member will be present. If you or your child get upset or cannot continue, we will give you support and help. If you or your child seems to be a risk to themselves or others, we will also help you. You should discuss the risk of being in this study with the study staff.

Taking part in this research study may involve providing information that you consider confidential or private. Efforts, such as coding research records, keeping research records secure and allowing only authorized people to have access to research records, will be made to keep your information safe.

Are There Benefits to Taking Part in the Study?
Being in this study may improve you and your child's quality of life. From being in the study, you and your child may learn things to do to deal with pain and the problems your child has because of pain. Your child may also see decreases in pain, and may be more active because of being in the study. Thus, the gains of the study seem to be much greater than the possible risks.

What Other Choices Are There?
This is not a treatment study. You and your child's alternative are not to participate in this study.

What about the Use, Disclosure and Confidentiality of Health Information?
By taking part in this research study, your child's personal health information, as well as information that directly identifies you and your child, may be used and disclosed. Information that identifies you and your child includes, but is not limited to, such things as you and your child's name, address, telephone number, and date of birth. Your child's personal health information includes all information about you and your child which is collected or created during the study for research purposes. It also includes your child's personal health information that is related to this study and that is maintained in your child's medical records at this institution and at other places such as other hospitals and clinics where you may have received medical care. Examples of your child's personal health information include your child's health history, your family health history, how you and your child respond to study activities or audiotapes and information from study visits, phone calls, surveys, and physical examinations.

Your child's personal health information and information that identifies you may be given to others during and after the study. This is for reasons such as to carry out the study, to determine the results of the study, to make sure the study is being done correctly, to provide required reports and to get approval for new products.

Some of the people, agencies and businesses that may receive and use your child's health information are the research sponsor; representatives of the sponsor assisting with the research; investigators at other sites who are assisting with the research; central laboratories, reading centers or analysis centers; the Institutional Review Board; representatives of Wake Forest University Health Sciences and North Carolina Baptist Hospital; representatives from government agencies such as the Food and Drug Administration (FDA), and the Department of Health and Human Services (DHHS).

Some of these people, agencies and businesses may further disclose your child's health information. If disclosed by them, your child's health information may no longer be covered by federal or state privacy regulations. Your child's health information may be disclosed if required by law. Your child's health information may be used to create information that does not directly identify you. This information may be used by other researchers. You and your child will not be directly identified in any publication or presentation that may result from this study unless there are photographs or recorded media which are identifiable.

If this research study involves the treatment of a medical condition, then information collected or created as part of the study may be placed in your child's medical record and discussed with individuals caring for your child who are not part of the study. This will help in providing your child with appropriate medical care. In addition, all or part of your child's research related health information may be used or disclosed for treatment, payment, or healthcare operations purposes related to providing your child with medical care.

When you sign this consent and authorization form you authorize or give permission for the use of your child's health information as described in the consent form. You or your child can revoke or take away your authorization to use and disclose your child's health information at any time. You do this by sending a written notice to the investigator in charge of the study at the following address:

Alexandra Boeving Allen, Ph.D.
Medical Center Boulevard
Winston-Salem, NC 27157

If you withdraw your authorization you will not be able to be in this study. If you withdraw your authorization, no new health information that identifies your child will be gathered after that date. Your child's health information that has already been gathered may still be used and disclosed to others. This would be done if it were necessary for the research to be reliable. You and your child will not have access to your health information that is included in the research study records until the end of the study.

This authorization does not expire.

What Are the Costs?
There are no costs to you for taking part in this study. All study costs, including procedures related directly to the study will be paid for by the study. Costs for you and your child's regular medical care, which are not related to this study, will be your own responsibility.

Will You Be Paid for Participating?
We will give your family $10 for the first visit. We will also give your family $10 for each visit that you and your child take part in. We will also give your family $20 for filling out the forms at 2-, 4-, and 6-months after the study visits end.

What Are My Rights as a Research Study Participant?
Taking part in this study is voluntary. You and your child may choose not to take part or may leave the study at any time. Refusing to participate or leaving the study will not result in any penalty or loss of benefits to which you or your child are entitled. If you decide to stop participating in the study we encourage you and your child to talk to the investigators or study staff first to learn about any potential health or safety consequences. The investigators also have the right to stop you or your child's participation in the study at any time. This could be because your child represent a risk to his/her self or others, requiring additional therapeutic intervention or because the entire study has been stopped.

Whom Do I Call if I Have Questions or Problems?

For questions about the study or in the event of a research-related injury, contact the study investigator, Alexandra Boeving Allen at 336-716-1284 (after hours please page at 336-806-6206).

The Institutional Review Board (IRB) is a group of people who review the research to protect your rights. If you have a question about your rights as a research participant, you should contact the Chairman of the IRB at (336) 716-4542.

You will be given a signed copy of this consent form.

Signatures
I agree to take part in this study. I authorize the use and disclosure of my child's health information as described in this consent and authorization form. If I have not already received a copy of the Privacy Notice, I may request one or one will be made available to me. I have had a chance to ask questions about being in this study and have those questions answered. By signing this consent and authorization form, I am not releasing or agreeing to release the investigator, the sponsor, the institution or its agents from liability for negligence.

_____
Subject Name (Printed)

_____ _____
Subject Signature Date

_____ _____
Person Obtaining Consent Date

I hereby acknowledge the above and give my voluntary consent for the participation of my child _____ (write in child's name) in this study. By signing this consent and authorization form, I am not releasing or agreeing to release the researchers, the institution or its agents from liability for negligence.

_____
Legally Authorized Representative Name (Print)

The above named Legally Authorized Representative has legal authority to act for the research subject based upon (specify health care power of attorney, spouse, parent, etc.)

_____
Relationship to the Subject

_____ _____
Legally Authorized Representative Signature Date

Appendix C

CBFT Session Checklists

Session One

For each component below, write a 1 in the space if the component was covered in the session. Write a 0 if the component was not covered.

Clinical interview to assess pain with parent and child/adolescent

___ a. Frequency

___ b. Duration

___ c. Location

___ d. Intensity

___ e. What does the child do when in pain (i.e., how cope)?

Rapport building

___ f. Built rapport

Educational Component

___ g. Rationale for CBT intervention

___ h. Encouraged to take belief that can handle/take control of pain

Concepts introduced

___ i. Breathing

___ j. Imagery – develop age appropriate/culturally sensitive scripts with child

___ k. Relaxation exercises

___ l. Rationale – connection between stress and pain

Summary and preparation for homework

___ m. Practice breathing and relaxation twice a day

___ n. Daily diaries

___ o. School attendance

___ p. Use of breathing, imagery, relaxation at school

Total Number of 1's for Session One: _____

Session Two

For each component below, write a 1 in the space if the component was covered in the session. Write a 0 if the component was not covered.

Diary Discussion

___ a. Review pain diaries

___ b. Triggers and consequences of pain

___ c. Relaxation techniques considered helpful?

___ d. Encouraged to take belief that can handle/take control of pain

Model/practice the following:

___ e. Breathing

___ f. Imagery

___ g. Relaxation exercises

Self-Talk

___ h. Introduce self-talk

___ i. Challenging negative thoughts and recognize as false

Summary and preparation for homework

___ j. Continuing relaxation exercises

___ k. Negative and positive thoughts tell self

___ l. Daily diaries

Total Number of 1's for Session Two: _____

Session Three

For each component below, write a 1 in the space if the component was covered in the session. Write a 0 if the component was not covered.

Diary Discussion

___ a. Review pain diaries

___ b. Relaxation exercises

___ c. Review negative self-talk

___ d. Positive self-statements to replace negative statements

___ e. Encouraged to take belief that can handle/take control of pain

Focus on cognitive components of pain

___ f. Introduce snowballing

___ g. Introduce catastrophizing

___ h. Importance of distraction (activities)

___ i. Demonstrate/role-play activity in session

Summary and preparation for homework

___ j. Relaxation exercise

___ k. Snowballing and positive self-statements

___ l. Distraction

___ m. Daily diaries

Total Number of 1's for Session Three: _____

Session Four

For each component below, write a 1 in the space if the component was covered in the session. Write a 0 if the component was not covered.

Have child demonstrate knowledge to parent

___ a. Have child complete a cartoon – indicating know what to do when pain occurs

Diary Discussion

___ b. Review pain diaries

___ c. Relaxation exercises

___ d. Stop snowballing, using positive self-statements

___ e. Use of distraction

Reframe role of parent from protector to coach

___ f. Encourage behavior incompatible with being sick

___ g. Praise well-behavior

___ h. Limiting discussion of pain episodes

___ i. Encourage use of coping (cognitive) and control (relaxation) techniques

___ j. Discussed: What are the parents' typical coping strategies?

___ k. Limit secondary gain from sick behavior

Summary and preparation for homework

___ l. Relaxation

___ m. Use of cognitive techniques

___ n. Distraction

___ o. Daily diaries

Total Number of 1's for Session Four: _____

Session Five

For each component below, write a 1 in the space if the component was covered in the session. Write a 0 if the component was not covered.

Diary Discussion

___ a. Review pain diaries and homework

Review Cognitive and Behavioral Strategies Introduced in treatment

___ b. Relaxation

___ c. Cognitive techniques

___ d. Distraction tools

___ e. Does the child know how to use the strategies implemented?

Recognize/summarize

___ f. Progress

___ g. Hard work

___ h. Position of control over pain

Total Number of 1's for Session Five: _____