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THE IMPACT OF SICKLE CELL DISEASE ON THE FAMILY: AN EXAMINATION OF THE ILLNESS INTRUSIVENESS FRAMEWORK

by

JOSIE S. WELKOM

Under the Direction of Dr. Lindsey Cohen

ABSTRACT

Sickle Cell Disease (SCD) is a genetic disorder that affects approximately 1 out of every 600 African-American newborns (NHLBI, 2006). SCD and its associated symptoms can have widespread impact on both the psychological functioning of the individual diagnosed with the illness and their families. The purpose of this study was to apply the illness intrusiveness framework to better understand the relations among vaso-occlusive pain crises (VOC), child age, pediatric health related quality of life (QOL), and parental psychosocial adjustment. Participants included 103 parent-child dyads. Parents completed a background form, the Brief Symptom Inventory-18, and the Illness Intrusiveness Rating Scale. Children completed the Pediatric Quality of Life Inventory. Results revealed that experiencing a greater frequency of VOC's was related to decrements in QOL across domains. However, this relation was not mediated by

parental perceived illness intrusiveness. Further, results revealed that the effect of frequency of vaso-occlusive pain crises in children with SCD on parental psychosocial maladjustment is mediated by parental illness intrusiveness, which is contingent upon child age.

INDEX WORDS: African Americans, Caregivers, Family, Illness intrusiveness, Parents, Quality of life, Psychosocial adjustment, Sickle cell disease

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by

JOSIE S. WELKOM

A Dissertation Submitted in Partial Fulfillment of the Requirements for the Degree of

Doctor of Philosophy

in the College of Arts and Sciences

Georgia State University

2012

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THE IMPACT OF SICKLE CELL DISEASE ON THE FAMILY: AN EXAMINATION OF THE ILLNESS INTRUSIVENESS FRAMEWORK

by

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August 2012

DEDICATION

I would like to dedicate this work to my mother, Lorna Welkom, and my father, Harris Welkom, for their endless support and unconditional love. As a testament to the sacrifices they have made and their devotion to our family, I am forever grateful.

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Introduction

Sickle Cell Disease Overview

Sickle Cell Disease (SCD) is one of the most common childhood-onset, single-gene disorders, affecting primarily people of African descent (Yanni, Grosse, Yang, & Olney, 2009). From an evolutionary perspective, in comparison to individuals with normal hemoglobin, the SCD trait provides a survival advantage in regions where malaria is prevalent (Allison, 1964; Aidoo, et al., 2002). The SCD gene causes an abnormality in the iron-rich protein hemoglobin that is responsible for carrying oxygen through the blood and giving blood its red color. The abnormal hemoglobin causes cells to become "sickle shaped" resulting in irregular blood flow (National Heart, Lung, and Blood Institute ([NHLBI], 2010). The red blood cells can stick and block the flow of blood to the limbs and organs resulting in pain, organ damage, and a low blood count. For one to inherit the disease, two copies of the sickle cell gene (i.e., one from each parent) must be transmitted to the offspring. Thus, children whose parents each carry the trait will have a 25% chance of inheriting the disease. Worldwide, approximately 300,000 infants are born with SCD (Modell & Darlison, 2008). In the United States, 1 out of every 500 African-American newborns and 1 out of every 36,000 Hispanic newborns is born with SCD. Overall, between 70,000 and 100,000 people or 1 out of 12 African Americans are currently living with the disease in the United States (NHLBI, 2010).The sickle cell trait is most common in individuals originating from the following countries or regions: Africa, South America, Central America, the Caribbean islands, the Mediterranean, India, and Saudi Arabia.

Due to advances in clinical recognition, newborn screening, and therapeutic and preventative interventions, the mortality rates of children with SCD have decreased by approximately 53% over the past four decades (Davis, Schoendorf, Gergen, and Moore, 1997;

Yanni et al., 2009). Given that children with SCD are living longer, the psychosocial impact of the disease on the patient and their families becomes a more significant issue. Specifically, both children with SCD and their caregivers are at risk for significant psychosocial maladjustment (for a review see, Edwards et al., 2005).

Symptoms of SCD include a vast array of physiological, neurocognitive, and psychological comorbidities (McClellan, Schatz, Sanchez, & Roberts, 2008; Midence & Shand, 1992; Schatz & McClellan, 2006; Shapiro, 1993). Physiological symptoms of SCD include vaso-occlusive pain crises, anemia, dactylitis or hand-foot syndrome, eye damage, splenic sequestration, decreased ability to fight infections, acute chest syndrome, delayed growth and puberty, leg ulcers, stroke (occurring in approximately 10% of children due in part to blood cells sticking to the walls of the blood vessels in the brain limiting blood flow), gallstones, damage to organs and/ or body tissue, and priapism (Centers for Disease Control and Prevention [CDC], 2011) .

Individuals with SCD, especially children during the first decade of life, are also at risk for the development of neurocognitive deficits (Brown et al., 1993; Chapar, 1988; Daly, Kral, & Brown, 2008; Fowler, Johnson, & Atkinson, 1985; Fowler et al., 1988; Swift et al., 1989). Despite the lack of apparent neurological symptoms, approximately 20-30% are at risk for silent strokes also defined as neurological changes in brain imaging (Kinney et al., 1999) and approximately 5-8% of children are at risk for overt strokes or cerebrovascular impairments (Balkaran et al., 1992; Ohene-Frempong et al., 1998). Transcranial Doppler ultrasonography (TCD) studies are conducted approximately once per year in children greater than 2 years of age to assess blood flow velocity in the middle cerebral or internal carotid artery which is associated with increased stroke risk (National Institutes of Health [NIH], 2002). Resulting cognitive

impairments may include decrements in overall intellectual functioning, language and verbal abilities, visual-motor and visual-spatial processing, memory, sustained attention, executive functions, and academic achievement (for a review, see Kral, Brown, & Hynd, 2001). The array of symptoms adds to the difficulties of parents raising and caring for children with SCD.

Sickle Cell Disease Pain

The hallmark feature of SCD is frequent and unpredictable pain, termed vaso-occlusive crises (VOC's; Shapiro & Ballas, 1994), which account for approximately 25% of hospital visits in children with SCD (Rees et al., 2003). VOC's are the result of "sickling" of the blood vessels leading to a lack of flexibility. Specifically, the red blood cells become sickle shaped and sticky, blocking blood flow through the vessels which results in pain, ischemia, and infarction (Ballas, 1998; Bookchin & Lew, 1996; Simon, Lobo, & Jackson, 1999). VOC's that result from blocked blood flow are defined as acute episodes, whereas VOC's resulting from damage due to repeated episodes and tissue ischemia are characterized as chronic (Franck, Treadwell, Jacob, & Vichinsky, 2002). The frequency of these pain episodes ranges from fewer than once a year to multiple times a day. Jacob et al. (2003) found pediatric sickle cell patients experienced pain an average of 4.5 (*SD* = 3.6) days prior to admission. In several diary studies with children and adolescents, VOC's ranged from 2.0-3.6 days within two to seven week periods (Dampier, Ely, Brodecki, & O'Neal, 2002; Gil et al., 2000; Maikler, Broome, Bailey, & Lea, 2001).

Treatment of SCD VOC's typically includes nonsteroidal anti-inflammatory drugs, opioids, and adjuvant medications (i.e., increases the analgesic effect of opioids, reduces the side effects, or manages associated symptoms; Benjamin et al., 1999). Transfusion therapy is utilized in patients with chronic and severe pain (Styles & Vichinsky, 1994). Several clinical trials have documented the effectiveness of hydroxyurea in decreasing the rate of painful episodes in adults

(for a review, see Bonds, 2005) and children (Ware, Zimmerman, & Schultz, 1999). Recent medical advances have identified allogenic bone marrow transplantation as a cure for sickle cell anemia (Walters et al., 1996). Research has consistently shown that between 60% and 90% of VOC's are managed at home by the patient or caregiver (Dampier et al., 2002; Fuggle, Shand, Gill, & Davies, 1996; Gil et al., 2000; Shapiro et al., 1995). However, analgesic treatments may not completely eliminate pain. For example, Jacob et al. (2005) found that of the 27 recruited pediatric patients with SCD presenting at an emergency room with vaso-occlusive pain, 44.4% reported that they did not experience pain relief from at-home analgesics. Alternatives to the analgesic therapies include physical strategies (e.g., application of heat, massages, and fluid intake) and psychosocial treatments (e.g., coping skills training, biofeedback, hypnosis, and relaxation techniques). Thus, SCD has been conceptualized as a disease with both psychosocial and physiological complications (Edwards et al., 2005). Given the unpredictable, frequent, and sometimes severe nature of SCD pain, coupled with its inconsistent response to intervention, SCD pain is a challenge for both clinicians and caregivers to manage (Gil et al., 1997; Waters & Thomas, 1995)

Psychological Impact of Sickle Cell Disease

SCD, and especially the pain episodes, can have widespread impact on both the psychological functioning of the individual diagnosed with the illness and their families. Children with SCD are at risk for maladjustment in almost every area of daily functioning (Barrett et al., 1988; Palermo, Riley, & Mitchell, 2008). Specifically, SCD has been associated with several indicators of psychological maladjustment including emotional and behavioral problems, poor self concept and interpersonal functioning, limited athletic abilities (due in part to illness restrictions), and poor academic performance (Eaton, Haye, Armstrong, Pegelow, &

Thomas, 1995; Morgan & Jackson, 1986; Noll, Reiter-Purtill, Vannatta, Gerhardt, & Short, 2007; Schaeffer, Gil, & Porter, 1999; Shapiro et al., 1995; Thompson et al., 1993).

With respect to the family, caregivers of children with SCD are burdened with missed work, increased family stress, and increased disease care demands, which is due in part to the unpredictability of pain crises care in SCD (Moskowitz et al., 2007). Caregivers of children with SCD are tasked with the responsibility for managing their child's care, which includes encouraging their child to engage in preventative behaviors, managing pain episodes, teaching coping skills, and providing adequate nutrition/ hydration. Moreover, the primary caregivers often report a lack of support by family and friends during the child's pain crises, which contribute to feelings of hopelessness, helplessness, and frustration (Midence, Fuggle, & Davies, 1993).

Health Related Quality of Life

Research has demonstrated that there is a unique interplay between the patient's psychosocial adjustment and the pathophysiology of SCD (Edwards et al., 2005). Given the increase in medical advancements and subsequent decreases in disease morbidity and mortality, more attention has focused on quality of life (QOL), which is an individual's assessment of his or her satisfaction with various aspects of his or her life (e.g., physical, emotional, school, social). Health related quality of life (HRQOL) refers more specifically to the impact of the child's illness on their subjective well-being (Hays, 1995; Pal, 1996). Measuring QOL has become increasingly important for its function in evaluating interventions, assessing prognostic factors, comparing therapies, and allocating resources (Panepinto, Pajewski, Foerster, Sabnis, & Hoffman, 2009).

Although there are few studies, findings suggest that QOL outcomes in children with SCD are generally poor (Kater et al., 1999; Palermo, Schwartz, Drotar, & McGowan, 2002; Panepinto, O'Mahar, DeBaun, Loberiza, & Scott, 2005). For example, Kater et al. (1999) found that a pediatric population with SCD had lower daily functioning and general physical limitations than parents of healthy children. In 2002, Palermo et al. found that children with SCD were experiencing more psychosocial maladjustment compared to the healthy controls. Moreover, children with SCD had significantly more limited general health and physical functioning, more limitations in their academic functioning and social activities attributed to their physical health, and more behavior and emotional problems when compared to a healthy control group. Moreover, in an adult SCD population, research has shown that the frequency of sickle cell pain episodes over a 12 month period was associated with impairment in QOL (Anie, Steptoe, & Bevan, 2002).

With approximately 10-20% of children reporting frequent and severe SCD-related pain (Walco & Dampier, 1990), it is likely that recurrent pain negatively impacts QOL. Research has shown that, SCD pain predicts several facets of QOL including school absences (Shapiro et al. 1995), lower academic performance (Hurtig, Koepke, & Park 1989), decreased participation in social activities (Fuggle et al., 1996), and sleep disruption (Dinges et al., 1990). Further, the frequency of these pain episodes is associated with decreases in QOL (Palermo et al., 2002; Panepinto et al., 2005)

Panepinto et al. (2009) examined the role of SES and disease severity in pediatric SCD. The criteria used to define disease severity included SCD-related complications (i.e., stroke, acute chest syndrome, hospitalizations for VOC's, and recurrent priapisms) and interventions (i.e., hydroxyurea or bone marrow transplantation). Panepinto et al. (2009) were interested in

clarifying the compounding nature of impoverishment in the African American community on reports of QOL. Participants included 104 children with SCD and 74 healthy controls aged 2-18 years. Results supported prior research documenting the negative effects of disease severity on QOL in children with SCD (Palermo et al., 2008; Panepinto et al., 2005; Panepinto, Pajewski, Foerster, & Hoffmann, 2008). Overall, children with the poorest QOL were those with a higher disease severity (e.g., frequent pain), older age, and more economically disadvantaged families. Given the debilitating effects of SCD pain, it is likely that the frequency with which children experience VOC's will greatly impact their QOL.

Chronological Age

Research has shown that developmental status has a significant influence on psychological adjustment in pediatric populations (Lavigne & Faier-Routman, 1992; Kell, Kleiwer, Erickson, & Ohene-Frempong, 1998). Previous research has found the relation between caregiver distress and child maladjustment in pediatric chronic illness to be interactional in nature (Chaney et al., 1997; Thompson, Gil, Burbach, Keith, & Kinney, 1993a, 1993b). Thus, it is important to consider the role of developmental status on caregiver and child adjustment. Adjustment to chronic illness is influenced by individual, family, and illness factors (Rolland & Williams, 2005). Typically, a gradual shift in disease-related responsibility from parent to child occurs over the course of development (La Greca, 1998; Shemesh et al., 2004) such that greater autonomy and independence are anticipated. This change in roles and responsibilities can actually lead to increases in both stress and conflict in both the adolescents and caregivers (Leonard, Garwick, & Adwan, 2005; Quittner, Espelage, Ievers-Landis, & Drotar, 2000). Research in other pediatric chronic illness populations has found that decreases in parental involvement is associated with poor disease management in adolescents with cystic fibrosis

(Zindani, Streetman, Streetman, & Nasr, 2006), diabetes (Anderson, Auslander, Jung, & Miller, 1990), and HIV (Mellins, Brackis-Cott, Dolezal, & Abrams, 2004). Therefore, older children with a chronic illness may actually require greater parental assistance and less autonomy (Holmbeck & Kendall, 2002) than healthy comparison adolescents. Caregiving demands which are inconsistent with normative development have been associated with greater distress especially among parents of older children (Andrews et al., 2009; Barakat, Patterson, Tarazi, & Ely, 2007). Over time, the prolonged stress of raising a chronically ill child may impact caregiver adjustment. Aside from the additive effect of attending to multiple stressors, it is likely that caregiver's expectations of their child's ability to meet age-related milestones will influence their subjective experiences. Of note, it is important to consider that whereas age may be indicative of developmental expectations, it is not synonymous with biological indices of child development. Thus, in light of the relation between parental age-related expectations and their own adjustment in other chronic illness populations, it will be important to explore the impact of developmental status in pediatric SCD.

Caregiver Adjustment

Caregivers of children with a chronic illness often report increased parental distress and negative affect (Drotar, 1997). Thus it is not surprising that caregivers of children with SCD have alarmingly high rates of maladjustment. For example, previous research has consistently found that approximately 24% of parents of young children with SCD report clinically significant levels of psychological distress; similar to that of adolescents with SCD (Thompson, Gustafson, Bonner, & Ware, 2002). In tandem, Brown et al. (2000) found that 35% of the primary caregivers in the sample met criteria for clinically significant levels of poor adjustment and 65% were at risk. The rates of psychosocial maladjustment in parents of children with SCD

are similar to the high rates found in individuals diagnosed with a medical condition (20%-25%; American Psychiatric Association, 2005).

Bachanas et al. (2001) found that parental psychosocial maladjustment was a significant predictor of their child's psychosocial maladjustment. Research has shown that the general outcome for caregivers of a child with a chronic illness is similar to that of the child (Timko, Stovel, & Moos, 1992). Moskowitz et al. (2007) found that in addition to time providing crisis care, parents of children with SCD are spending a greater proportion of time in disease-related technical care such as medication administration and diagnostic procedures than parents of a child with HIV. Furthermore, possibly due to the unpredictability of pain crises, caregivers of children with SCD are also reporting greater care burden (Moskowitz et al., 2007). Fifty percent of the caregivers of children with SCD in this study were at risk for clinical depression compared to 34% of parents of children with HIV.

Although there is limited research examining the relation between SCD pain and parental psychosocial adjustment, there is ample research documenting this relation in pediatric chronic pain (for a review see Palermo & Eccleston, 2009). Given that SCD is a chronic illness characterized by both acute and chronic pain, it is likely that the chronic pain literature can be applied to the population under study. Though rare, as of late more attention has been directed towards the families of children experiencing pain (Palermo & Chambers, 2005). Specifically, parents of children with chronic pain are not only directly impacted by the stress of parenting a chronically ill child, they are influential in both the child's adjustment to and experience of the pain (Palermo & Eccelston, 2009). Consistent with the research on parents of children with SCD, research with parents of children with chronic pain has shown that these parents often report high

levels of anxiety and depression (Campo et al., 2007; Eccleston, Crombez, Scotford, Clinch, & Connell, 2004; Jordan, Eccleston, & Crombez, 2008).

Jordan, Eccleston, and Osborn (2007) utilized qualitative methodology to examine the caregiver's experience of parenting an adolescent with chronic pain. Jordan et al. (2007) conducted focus groups with 17 parents of adolescents (age 11-18 years) with chronic pain recruited from two tertiary care clinics in the United Kingdom. The researchers utilized Interpretive Phenomenological Analysis (IPA), a qualitative analytical method, for exploring the data. Two themes emerged which were labeled the caregiver's "struggle for control and coherence" and a "very different life". Moreover, these caregivers highlighted the disruption in developmental milestones which contributed to their feelings of uncertainty. Though there are disease-related differences, these themes parallel the difficulties regarding unpredictability which is associated with SCD VOC's.

Palermo and Chambers (2005) examined the role of parent and family factors associated with pediatric chronic pain and disability. Palermo and Chambers (2005) argue that there has been a lack of an integrative contextual framework for adequately conceptualizing the effects of parenting-specific behaviors and broader family factors on child pain. Their model highlights the reciprocal relationship between child pain and parental factors (e.g., psychosocial adjustment). Palermo and Chambers (2005) suggest that researchers design future studies based on integrative theoretical models which will contribute to our understanding of the multifaceted nature of pediatric pain.

The Illness Intrusiveness Framework

The Illness Intrusiveness Framework (Devins et al., 1983) provides a model for explaining how disease factors (e.g., vaso-occlusive pain episodes) might impact the patient

(e.g., QOL) and their caregivers (e.g., parent adjustment). Illness intrusiveness refers to when disease-related factors (e.g., pain) interfere with valued activities and interests. This theory postulates that disease (e.g., pain) and treatment factors (e.g., disruptive treatment schedules) influence subjective well-being (e.g., quality of life and psychological distress) indirectly through their effects on illness intrusiveness (Devins, 2010; see Figure 1). Moreover, illness intrusiveness is a cognitive appraisal mechanism, which indirectly compromises psychological adjustment through reducing both perceived control and the resulting positive outcomes associated with participation in valued activities (Devins, 1994; Devins, Seland, Klein, Edworthy, & Saary, 1993). The illness intrusiveness framework also takes into account psychological, social, and contextual factors. These factors are hypothesized to contribute directly and indirectly to subjective well-being. Specifically, stable factors such as gender and cultural heritage are thought to moderate the relation between disease/treatment factors on illness intrusiveness and state-specific factors such as developmental stage and stigma are thought to moderate the relation between illness intrusiveness and psychological adjustment (Devins, 2010; see Figure 1).

There is an extensive body of literature supporting the illness intrusiveness theoretical framework in over 36 chronic disease populations including cancer, heart disease, HIV, end-stage renal disease (ESRD), multiple sclerosis, non-insulin dependent diabetes mellitus, lupus, and transplant (e.g., heart, liver, lung, renal; for a review, see Devins, 2010). Originally, Devins et al. (1983) developed the Illness Intrusiveness Rating Scale (IIRS) to assess this construct in ESRD patients. However, it has been successfully adapted for its use with pediatric patients (Wagner et al., 2003) and their parents (Andrew et al., 2009). The IIRS assesses the following three life domains: Relationships and Personal Development (e.g., family/ social relations, self-

expression and improvement, religious expression, community and civic involvement, passive recreation), Intimacy (e.g., relationship with partner, sex life), and Instrumental Life (e.g., health, work, financial situation, active recreation; Devins et al., 2001). To date, the illness intrusiveness theoretical framework has not been examined in pediatric SCD. However, studies of illness intrusiveness in other medical populations might illuminate how this framework would apply to children with SCD and their parents.

Devins, Bezjak, Mah, Loblaw, and Gotowiec (2006) evaluated the moderating effects of contextual factors (e.g., age, education, income, stressful life events) on the experience of illness intrusiveness in an adult cancer population. Results indicated that younger, lower income, and patients experiencing at least one or more stressful life events reported higher illness intrusiveness scores. Moreover, results revealed significant 2-way interactions among the Instrumental Life factor and each of the hypothesized contextual moderator variables such that younger, more highly educated, higher income, and patients who experienced at least one stressful life event differentially reported significantly higher illness intrusiveness ratings in the Instrumental Life domain as opposed to the Relationships and Personal Development and Intimacy domains. These results suggest that lifestyle disruptions across domains vary as a function of the context in which the illness is experienced.

Devins et al. (2009) further explored the moderating effects of age and culture (e.g., attitudes, values, and beliefs) on the relation between illness intrusiveness and psychological adjustment in adult patients with rheumatoid arthritis (RA). Clinical symptoms of RA include pain and stiffness, which progresses into increasing deformity and disability. Specifically, Devins et al. (2009) hypothesized that cultural variables influence the emotional impact of illness intrusiveness and this is differentially affected by age. This study used individualist (i.e.,

autonomy, personal goals, individual attitudes, and personally advantageous relationships) and collectivist (i.e., group belonging, social norms, group norms, and communal relationships) attitudes to describe culture (Triandis & Gelfand, 1998). Contextual variables included demographics and recent negative life events; disease-related variables included pain severity, functional disability, and duration of illness. The results of this study revealed that horizontal individualism (i.e., independent, expectation of equality) moderated the effects of illness intrusiveness on psychological distress in young and middle-aged adults with RA. Additionally, illness intrusiveness was associated with more psychological distress in younger and middle-age adults compared to older adults. This study provides evidence for culture and life-span developmental factors as moderators on the illness intrusiveness-psychological distress relation in adults with RA.

Talbot, Nouwen, Gingras, Belanger, and Audet (1999) examined the relations among diabetic complications, perceived control, illness intrusiveness, and depressive symptoms in 237 adults with diabetes. Specifically, Talbot et al. (1999) hypothesized that illness intrusiveness directly and indirectly (i.e., through perceived control) mediates the relation between disease-specific variables and depressive symptoms. The results of the study revealed that while controlling for the effects of the disease-related variables, the hypothesized model predicted 65% of the variance in depressive symptomatology. Moreover, it was found that whereas illness intrusiveness predicted 61% of the variance in depressive symptomotology, perceived control only contributed to approximately 4%. The finding of this study further supports illness intrusiveness as an integral factor in predicting psychosocial functioning in patients with a chronic illness.

With regards to the unpredictability of onset of symptoms, epilepsy is comparable to that of SCD. Poochikian-Sarkissian, Sidani, Wennberg, and Devins (2008) examined the illness intrusiveness theoretical framework and compared the impact of pharmacological and surgical treatments on illness intrusiveness and quality of life (QOL) in adults afflicted by epilepsy. A path analysis was conducted, which supported the tenets of the illness intrusiveness theoretical framework. Specifically, successful surgical treatments were associated with more positive illness intrusiveness and healthier psychological adjustment as compared to pharmacological treatment. Moreover, illness intrusiveness significantly mediated the relation between disease-related factors and psychological adjustment (i.e., depression, self-esteem, and QOL). These results suggest that improvements in psychological adjustment can be achieved through the development of biopsychosocial approaches geared towards reducing the illness-induced disruptions (i.e., illness intrusiveness) in lifestyle and increasing the positive outcomes associated with participation in valued activities.

Given the extensive literature linking parental adjustment to child health outcomes (Chaney et al., 1997; Dahlquist et al., 1993; Thompson et al, 1993a, 1993b), Andrews et al. (2009) sought to examine age-related patterns among parent-reported illness intrusiveness and parental distress in caregivers of children with juvenile rheumatoid diseases (JRDs). In light of parental expectations for increased independence and autonomy paralleling child age, it is likely that illness-induced disruptions may be particularly distressing for parents when these expectations are not achieved, such that older children are not meeting their developmental expectations. Specifically, Andrews et al. (2009) hypothesized that child age would moderate the illness intrusiveness-parental psychological adjustment with parents of older children reporting more psychological maladjustment than parents of younger children. There was a significant

main effect for illness intrusiveness on parental psychological adjustment, and an illness intrusiveness X age interaction was found. Specifically, consistent with the hypothesis, parental perceived illness intrusiveness was more closely associated with parental psychological adjustment as child age increased. Post-hoc probes revealed that whereas parental illness intrusiveness was significantly associated with parental psychological maladjustment in older children, this relation was reduced in parents of younger children. These findings suggest that when parents' normal developmental expectations, such as increased autonomy and independence, are not met or are disrupted, parents experience more psychological distress possibly due to the additive effects of stress (Streisand, Kazak, & Tercyak, 2003). The family life cycle developmental framework provides evidence that parents may perceive atypical child development in the context of chronic illness more distressing because of the increased burden (Rolland, 1987).

Wagner et al. (2003) examined the moderating effects of illness intrusiveness on the parent distress-child depressive symptoms relation in a JRD population. A multivariate transactional stress and coping model (Thompson et al., 1993) was utilized to analyze the direct effects of parent distress on child depressive symptoms and to examine the role of children's perceived illness intrusiveness on the parent-child adjustment relation. Participants included 45 children between age 9 and 17 diagnosed with JRD and their parents. Results revealed that while controlling for demographic and disease-related variables, the interaction of parent distress and child illness intrusiveness was significant and contributed 5.3% of unique variance in child depressive symptoms. Specifically, post hoc analyses suggest that these results were magnified under conditions of high perceived child illness intrusiveness and not apparent under conditions of low perceived child illness intrusiveness. Thus, parent distress was only predictive of child

depressive symptoms when children reported high perceived illness intrusiveness. This study provides further support for the transactional nature of parent-child adjustment in pediatric chronic illness.

Andrews et al. (2007) explored the role of racial group differences (Native American and Caucasian) on the relation between cognitive appraisals of illness intrusiveness on adjustment in parents of children with JRD. As of date, this is the only study to examine the relation between parents' perceived illness intrusiveness of their child's illness on their own psychological adjustment. It should be noted that both illness intrusiveness (Devins, et al., 1992; Wagner, et al., 2003) and culture (Devins & Edworthy, 2000) have been implicated as factors critical to adjustment in chronic illness populations. Andrews et al. (2007) found that parental racial group membership moderated the illness intrusiveness-adjustment relation such that Native American parents who reported higher illness intrusiveness, similarly reported higher psychological maladjustment in comparison to Caucasian parents. Moreover, illness intrusiveness was not a significant predictor of psychological adjustment in Caucasian parents of children with JRD. This study provides further evidence for culturally-sensitive examinations of the cognitive appraisal-psychological adjustment relation in various illness populations.

Aims and Hypotheses

The purpose of the current study was to apply the illness intrusiveness framework to better understand the relations among vaso-occlusive pain crises (VOC's), child age, pediatric health related quality of life (QOL), and parental psychosocial adjustment.

Specific Aim 1 and Hypothesis. This study examined the potential mediating effects of parental illness intrusiveness on the VOC-QOL relation. Grounded in prior data showing that the relation between parental and child psychosocial outcomes are transactional in nature (Bachanas

et al., 2007; Chaney et al., 1997; Dahlquist et al., 1993; Thompson et al, 1993a, 1993b) in addition to the indirect effects of disease related factors on QOL through its effect on illness intrusiveness (Devins, 2009), it was hypothesized that the frequency of VOC's over the course of one year will influence subjective well-being of children (i.e., QOL) indirectly through its effect on parental illness intrusiveness (see Figure 2).

Specific Aim 2 and Hypothesis. This study examined the potential mediating effects of parental illness intrusiveness on theVOC-parental psychosocial adjustment relation. Based on findings that parent illness intrusiveness was related to distress in caregivers of children with juvenile rheumatic disease (Andrews et al., 2009; Wagner et al., 2003), it was hypothesized that the frequency of VOC's over the course of one year would influence parents' psychosocial adjustment indirectly through its impact on parents' illness intrusiveness (see Figure 3).

Specific Aim 3 and Hypothesis. Moreover, this study sought to explore age-related patterns of association among the aforementioned relations. Prior research has found that the psychosocial impact of illness intrusiveness is moderated by age in adult chronic illness populations (Devins et al., 2009; Devins, Beanlands, Mandin, & Paul, 1997; Devins et al., 1992). Moreover, child age has been shown to moderate the illness intrusiveness-parent distress relation (Andrews et al., 2009; Wagner et al., 2003). Specifically, it was hypothesized that child age would moderate the parent illness intrusiveness-parent psychosocial adjustment relation such that increased illness intrusiveness will be related to poorer parental psychosocial adjustment in parents of older children in comparison to parents of younger children (see Figure 4).

Method

Participants

Participants included 103 caregivers, ranging in age from 28 to 68 years of age (*M* = 41.1, *SD* = 8.04) presenting with their child at a regularly scheduled pediatric SCD appointment in the southeastern United States between May 2008 and August 2008. Based on a medium effect size of 0.15, sample size of 103, error probability of 0.05, and three predictors, a post hoc power analysis revealed that the study contained sufficient power of 0.91 (Faul & Erdfelder, Buchner, & Lang 2009). The majority of children were accompanied by their mother (*n* = 86, 83.5%) and the remaining children were accompanied by either their father (*n* = 9, 8.5%), a grandparent (*n* = 6, 5.8%), or a legal guardian identified as "other" (*n* = 2, 1.9%). One-hundred (97.1%) of the caregivers identified as "Black or African American", 1 (1.0%) identified as "Native Hawaiian or Pacific Islander", and 2 (1.9%) caregivers did not report their race. With regards to ethnicity, 1 (1.0%) caregiver identified as "Hispanic or Latino" and 9 (8.7%) did not report. The average years of education were 13.86 (*SD* = 2.06). Fifteen (14.5%) of caregivers reported an annual income of below $20,000, 37 (36%) ranged between $20,001 and 50,000, 23 (22.4%) ranged between $50,001 and $80,000, and 19 (18.4%) reported annual income greater than $80,000. Two (1.9%) caregivers failed to report their education level and 9 (8.7%) did not report income. The majority (*n* = 58) of caregivers were married (56.3%), 24 were single (23.3%), 16 were divorced (15.5%), 3 were separated (2.9%), and 2 (1.9%) did not report. Twenty-six (26.2%) caregivers reported having a chronic illness, most common of which included asthma (*n* = 9, 8.8%), sickle cell disease (*n* = 9, 8.8%), and diabetes (*n* = 3, 2.9%). Seven caregivers (6.8%) reported having a psychosocial disorder, which included anxiety (*n* = 4, 3.9%), depression (*n* = 3, 2.9%), and bipolar disorder (*n* = 1, 1.0%; see Table 1).

The sample of children whom the caregivers accompanied to the medical visit ranged in age from 8 to 18 years (*M* = 12.88, *SD* = 3.09). Fifty-six (54.4%) of the children were female and forty-seven (45.6%) were male. One-hundred (97.1%) of the children were "Black or African American", 1 (1.0%) was "Asian", and 2 (2.9%) caregivers failed to report their child's race. Eighty-eight (85.4%) of the children were not "Hispanic or Latino" and 15 (14.6%) failed to report. The most common SCD type was HbSS (*n* = 69, 67%), 13 (12.6%) had HbSC, 6 (5.8%) had HbSβ, and 15 (14.5%) did not specify a subtype. Thirty-four of the children had a co-existing chronic illness, most common of which was asthma (*n* = 31, 30.3%), 3 had heart murmurs (3.0%), and 2 had a history of acute chest syndrome (2.0%). Five (4.9%) children were diagnosed with Attention Deficit Hyperactivity Disorder (ADHD), 1 (1.0%) with anxiety, 1 (1.0%) with depression, and 1 (1.0%) with bipolar disorder. Most commonly, 1 (*n* = 33; 32.0%) additional sibling lived in the home, followed by 0 (*n* = 29; 28.2%) siblings, 2 (*n* = 24; 23.3%) siblings, 3 siblings (*n* = 9; 8.7%) or 4 or more (*n* = 4; 3.9%) siblings; 4 (3.9%) caregivers failed to report how many additional children lived in the home. Nine (8.7%) caregivers reported having at least 2 children in the home with SCD and 16 (15.5%) failed to report (see Table 1).

Of the 115 caregivers approached to participate in the study, 11 declined participation. Five caregivers reported not being interested in research, two caregivers identified time as their reason for decline, one did not feel comfortable due to current legal involvement, one father did not feel comfortable participating without the permission of the child's mother, one child was not officially diagnosed with SCD, and one male child was not interested but did not state his reason. In addition, one caregiver agreed to participate but reported that English was her second language and required considerable assistance completing the forms, thus she was eliminated from the final sample resulting in a final sample size of 103 parent-child dyads.

Measures

Background Information. Caregivers completed the Background Information Form. Questions assessed background information about the parent (e.g., relation to child, gender, age, ethnicity, race, education, occupation, family income, health status) and child (e.g., gender, age, ethnicity, race, and health status). These data will be used to describe the sample and child age will be used as a proxy for developmental status.

Sickle Cell Pain. Parents reported the total number of vaso-occlusive pain crises (VOC's) experienced in the past 12 months. Frequency of VOC's in pediatric SCD has been associated with decrements in QOL in children (Palermo, Schwartz, Drotar, & McGowan, 2002; Panepinto et al., 2005) and psychosocial adjustment in caregivers (Midence et al., 1993).

Health Related Quality of Life. The Pediatric Quality of Life Inventory (PedsQL; Varni, Seid, & Kurtin, 2001) was completed by children to assess their health related quality of life. The PedsQL is a 23-item generic quality of life measure designed for children and adolescents between 2 and 18 years of age. For the purposes of the current study, the PedsQL Child Report (ages 8-12) and the PedsQL Teen Report (ages 13-18), were utilized. The PedsQL assesses several domains of functioning, including Physical (8 items), Emotional (5 items), Social (5 items), and School (5 items) and utilizes a 5-point likert scale ranging from 0 (*never a problem*) to 4 (*almost always a problem*). As indicated by the scoring manual, raw scores on the PedsQL were transformed to scores ranging from 0 to 100, with higher scores representing higher quality of life. In addition to having specific scaled scores, the items on the PedsQL are averaged to yield a Total Scale Score and two summary scores, the Physical Health Summary Score and the Psychosocial Health Summary Score. In youth with SCD, whereas the PedsQL demonstrated adequate internal consistency at the level of the Total Score (Cronbach's alphas ranging from

0.86 – 0.92), discriminant validity across domains was variable due in part to younger children (i.e., <13) providing lower internal consistency scores (McClellan, Schatz, Sanchez, & Roberts, 2008). Within this sample, the PedsQL Total score was found to be internally consistent for the Child and Teen Reports (Cronbach's alphas ranged from 0.90 to 0.92). The PedsQL is considered a "well-established" instrument (Palermo et al., 2008). For the purposes of this study, the PedsQL Total Score will be used in primary analyses.

Caregiver Adjustment. The Brief Symptom Inventory-18 (BSI-18; Derogatis, 2000) was completed by caregivers to assess psychosocial adjustment. The BSI-18 is an 18-item psychosocial functioning measure written at a sixth grade reading level designed for adults eighteen years and older. The BSI-18 measures three scales (depression, anxiety, somatization) and one index (global severity). It utilizes a 5-point Likert scale (0 = *not at all* to 4 = *extremely*) to measure the amount by which certain problems have caused distress in the past seven days. The global severity index was used for primary analyses in the current proposal, and the subscales were analyzed for descriptive and exploratory purposes. The BSI-18 was designed for both primary care and community populations and has demonstrated acceptable internal consistency (Cronbachs's alphas ranging from 0.71 – 0.85; Derogatis, 1993). Within this sample, Cronbach's alpha was 0.92, suggesting strong internal consistency. Moreover, The BSI has been shown to be both reliable (Mullins et al., 1991, 1995) and valid (Derogatis, 1993) in prior studies. The GSI will be used for primary analyses in the current study.

Illness Intrusiveness. The Illness Intrusiveness Rating Scale (IIRS; Devins et al., 1983) was completed by parents to rate the extent to which their child's illness and/or treatment interferes with certain life domains. The IIRS is a 13-item self-report instrument which utilizes a 7-point likert scale ranging from 1 (*not very much*) to 7 (*very much*). The IIRS generates 3 –

Relationships and Personal Development (6 items), Intimacy (2 items) and Instrumental (4 items) – and a Total Score (13 items). The scores are created by averaging across the items within each subscale. Higher scores reflect higher illness intrusiveness. The IIRS utilized in the present study was slightly modified from the original version from *your illness* to *your child's illness* (Andrews et al., 2009). The original IIRS which utilized self- versus parent-report was found to be internally consistent (Cronbach's alpha = 0.91) and reliable in adult illness populations (0.79 – 0.85; Devins et al., 1990; Devins et al., 1993). In the one previous study utilizing the adapted parent version of the IIRS, Cronbach's alpha was 0.92 (Andrews et al., 2009). Within this sample, the adapted Illness Intrusiveness Scale-Parent version Total score was also found to be internally consistent (Cronbach's alpha = 0.95). For the purposes of this study, the IIRS-P Total score will be used for primary analyses.

Procedures

Families with a child presenting at an SCD clinic at two urban children's hospitals in the metro Atlanta area were informed about the study by clinic personnel and directed to receive additional information from a nearby researcher. The research assistant further explained the study and obtained consent if the caregiver was interested in participating. Families presented for one of four services at the clinics: a standard SCD appointment (47.6%), a standard SCD appointment with a pulmonary clinic (35.0%), a pain clinic (2.9%), or for infusion treatment (14.6%).

Before meeting with the physician, the caregiver was individually administered the measures (Background Information Form, IIRS-P, and BSI-18) and the child was individually administered the PedsQL in the waiting room. If the participants were unable to complete the measures prior to the child's clinic visit, they were encouraged to complete them during

additional wait time while in the physician's office (e.g., while waiting between the nurse and physician visits).

Data Analyses Overview

Preliminary analyses were conducted to characterize the sample, determine if covariates should be taken into consideration in primary analyses, and examine associations among the study variables. Primary data analyses were conducted as a series of regression models utilizing macros developed to test mediation (Preacher & Hayes, 2008), moderation (Hayes & Matthes, 2009), and mediated-moderation (Preacher, Rucker, & Hayes, 2007). Specifically, simple mediation models were conducted to examine the direct effect of pain on the outcome variables (i.e., pediatric quality of life and parental psychosocial adjustment) and the indirect effect of pain on the outcome variables through illness intrusiveness. Multiple regression analyses were conducted to examine the main effects of parental illness intrusiveness and child age and the respective interaction effect on parental psychosocial adjustment. Moreover, mediated-moderation analyses were conducted to examine the moderating role of child age on the indirect effect of pain on parental adjustment through illness intrusiveness. Bootstrapping, a nonparametric resampling procedure, was utilized to estimate the indirect effects and construct confidence intervals (Bollen & Stine, 1990; MacKinnon, Lockwood, & Williams, 2004; Preacher & Hayes, 2004; Shrout & Bolger, 2002).

Preliminary Analyses

Pearson's correlation revealed a significant positive correlation between child health-related quality of life and the presence of comorbid child psychiatric diagnoses, $r = .20$, $p = .05$. Significant negative correlations were also found between parental psychosocial adjustment and relationship to child (i.e., mother, father, grandparent, other) $r = -.22$ $p = .03$, parent age, $r = -.22$,

p = .03, and family income, r = -.21, p = .04. Analyses of Variance (ANOVAs) revealed that there were site differences in pediatric health related quality of life, F (1, 97) = 5.68, p = .02. Follow-up t tests suggested that on average, children at Site 1 reported significantly greater health-related quality of life (M = 12.92, SD = 2.47) than children at Site 2 (M = 74.37, SD = 14.47), t (96) = 2.38, p = 0.02, medium sized effect, d = 0.48 (Cohen, 1988). As such, these variables were controlled for in the primary analyses.

Assumptions for regression analyses (i.e., linearity, independence, homoscedasticity, and normality of errors) were tested and satisfied. The Durbin-Watson statistic was within the acceptable range of between 1.4 and 2.6 satisfying the assumption of independence of errors. Assumptions of homoscedasticity and normality of the error distributions was satisfied such that the residuals were normally distributed across levels of the predictor. Bootstrapping procedures do not require assumptions about the shape of the statistic sampling distribution when conducting inferential tests. Assumptions of single-mediator analyses (i.e., no misspecification of causal order, causal direction, unmeasured variables, or imperfect measurement) are often untestable but can be guided by experimental studies, theory, and qualitative methods (McKinnon, Fairchild, & Fritz, 2007). With regards to missing data, the aforementioned macros implement listwise deletion for cases with any missing data on variables within the model.

Primary Analyses

Model 1. Specific Aim 1 examined the indirect effect of pain (VOC; M = 3.55, SD = 4.20) on pediatric health-related quality of life (QOL; M = 70.61, SD = 16.33) through parent illness intrusiveness (IIRS-P; M = 31.42, SD = 20.09) utilizing single mediator analyses. Nonparametric bootstrapping analyses (Preacher & Hayes, 2004; Preacher et al., 2007) were utilized to test the mediational model. Results based on 10,000 bootstrapped samples indicated

that the total effect of the frequency of vaso-occlusive pain crises on pediatric health related quality of life was significant, β = -.92, *p* = .03, such that children with SCD whom experienced a greater frequency of VOC's reported poorer quality of life than those who experienced fewer VOC's. Moreover, whereas the frequency of VOC's did not have a not have a statistically significant direct effect on parent illness intrusiveness, β = .83, *p* = .13, the direct effect of parent illness intrusiveness on pediatric quality of life, β = -.17, *p* = .052, was approaching significance such that the greater the overall extent to which parent's reported that their child's illness interferes with certain life domains was associated with decrements in child reported pediatric quality of life. However, results revealed that the specific indirect path of VOC's on QOL through parent illness intrusiveness was not significantly different from zero ($95\%\ CI_{lower}$ = -.8529, $95\%\ CI_{upper}$ = .1297), indicating a nonsignificant mediator path. Mediation analyses based on 10,000 bootstrapped intervals using bias corrected and accelerated 95% confidence intervals showed that controlling for the effect of the covariates (presence of a comorbid psychiatric diagnosis, β = 9.29, *p* = .21; site differences, β = -6.20, *p* = .07), the IV (frequency of VOC's) had a nonsignificant total effect on the DV (QOL; β = -.17, *p* = .052), a nonsignificant residual direct effect (β = -.78, *p* = .06), and a nonsignificant indirect effect (β = -.14, *SE* = .15, CI_{lower} = -..8526, CI_{upper} =.1348; see Figure 2 and Table 3).

Model 2. Specific Aim 2 examine the indirect effect of pain (VOC; *M* = 3.55, *SD* = 4.19) on parental psychosocial adjustment (GSI; *M* = 6.88, *SD* = 9.24) through parent illness intrusiveness (IIRS-P; *M* = 31.42, *SD* = 20.09) utilizing single mediator analyses. Nonparametric bootstrapping analyses (Preacher & Hayes, 2004; Preacher et al., 2007) were utilized to test the mediational model. Results based on 10,000 bootstrapped samples indicated that the total effect of the frequency of vaso-occlusive pain crises on parental psychosocial adjustment was not

significant, β = .33, *p* = .16. Moreover, whereas the frequency of VOC's on parent illness intrusiveness was not significant, β = .77, *p* = .15, the direct effect of parent illness intrusiveness on parental psychosocial adjustment was approaching significance, β = 1.01, *p* = .06, such that the greater the overall extent to which parents' reported that their child's illness interferes with certain life domains predicted increases in parental maladjustment. However, results revealed that the specific indirect path of VOC's on QOL through parent illness intrusiveness was not significantly different from zero ($95\%\ CI_{lower}$ = -.0558, $95\%\ CI_{upper}$ = .9703), indicating a nonsignificant mediator path. Mediation analyses based on 10,000 bootstrapped intervals using bias corrected and accelerated 95% confidence intervals showed that controlling for the effect of the covariates (relation to child, β = -2.15, *p* = .13; parent age, β = -.10, *p* = .40; family income, β = .03, *p* = .92), the IV (frequency of VOC's) had a nonsignificant total effect on the DV (GSI; β = .33, *p* = .16), a nonsignificant residual direct effect (β = .17, *p* = .45), and a nonsignificant indirect effect (β = .16, *SE* = .18, CI_{lower} = -.0558, CI_{upper} = .9703; see Figure 3 and Table 4).

Model 3. Specific Aim 3 examined the moderating role of child age on the parent illness intrusiveness (IIRS-P) and parent psychosocial adjustment (GSI) relation. Both the independent and moderator variables were mean-centered prior to computing the interaction term to reduce multicollinearity. Hayes and Matthes (2009) outline computational procedures for estimating and probing interactions in OLS regression models. Specifically, regression coefficients, conditional effects, and regions of significance were generated. Results revealed that the model which included the covariates (i.e., relation to child, parent age, and family income), the focal predictor variable (parent illness intrusiveness) and the moderator variable (child age) significantly predicted parental psychosocial adjustment, F (6, 71) = 4.73, *p* = .0004. However, whereas there was a statistically significant main effect of parent illness intrusiveness on parental psychosocial

adjustment, b = .17, p = .0005, there was not a significant main effect for child age, b = -.03, p = .91. Most importantly, results revealed a significant interaction effect, thus the main effects will not be further interpreted. The interaction between parent illness intrusiveness and child age was significant, b = -.04, p = .03 suggesting that the relationship between parent illness intrusiveness and parental psychosocial adjustment is contingent on the age of the child. Post-hoc probing revealed that the interaction effect was significant at both low (1 *SD* below the mean), b = .30, p = .0001, and moderate (mean), b = .17, p = .0005, values of child age. The interaction was not significant at high values of child age (1 *SD* above the mean), b = .04, p = .54. Thus, parents of younger children are more likely to report that increases in the intrusiveness of their child's illness and/ or treatment on certain life domains are similarly associated with relative increases in their psychosocial maladjustment (see Figure 4 and Table 5).

Model 4. Given the significant moderator effect of child age (Specific Aim 3), model 4 utilized mediated moderation analyses (Preacher, Rucker, & Hayes, 2007) to examine whether parent illness intrusiveness (IIRS-P) would mediate the relationship between the frequency of vaso-occlusive pain crises (VOC) and parent psychosocial adjustment (GSI; Specific Aim 2), but only at low and moderate levels of child age (Specific Aim 3; see figure 6). Regression analyses revealed that the overall relationship between VOC's and GSI through IIRS-P was significant across child age, β = -.05, p = .008. However, the 95% confidence interval for the complete indirect effect was found to exclude zero for children at moderate age (M =12.77, SD = 3.11; β = .16, SE = .10, CI_{lower} = .0117, CI_{upper} = .6586) and at 1 SD below the mean age (β = .30, SE = .18, CI_{lower} = .0253, CI_{upper} = 1.3314) with 10,000 resamples. In contrast, the 95% confidence interval for children at 1 SD above the mean includes zero (β = .01, SE = .08, CI_{lower} = -.1536, CI_{upper} = .1718) with 10,000 resamples. Thus, these results suggest that the effect of frequency of

vaso-occlusive pain crises in children with SCD on parental psychosocial maladjustment is mediated by parental illness intrusiveness in parents of children at and/ or below the mean age of 12.77 (*SD* = 3.11) years within this sample (see Figure 5 and Table 6).

Discussion

The aim of this study was to examine vaso-occlusive crises, child age, parental illness intrusiveness, pediatric health-related quality of life, and parental psychosocial adjustment in children and parents of children with Sickle Cell Disease (SCD). Similar to other populations of parents of children with SCD in the literature (Barakat et al., 2007; Barakat, Lutz, Smith-Whitley, & Ohene-Frempong, 2005), the current sample of parents included mostly mothers who were in their early 40s and of middle-class income. In contrast to comparison samples (Brown et al., 2000; Barbarin, Whitten, Bond, & Conner-Warren, 1999, Kaslow et al., 1997), the current sample had a higher percentage of married parents and reported a higher annual income. Overall, parents reported relatively healthy psychosocial functioning (Derogatis, 2000; see table 1) which is inconsistent with previous research, documenting that caregivers of children with SCD are at risk for significant psychosocial maladjustment (Thompson et al., 2002; Brown et al., 2000; Thompson et al., 1993). Although it might be that this sample might simply be better adjusted than samples in other studies, differences might reflect variability in the methods and/or demographics of the sample. Specifically, the Thompson, et al. (2002) sample consisted of caregivers of newborns and psychosocial functioning was measured with the longer Brief Symptom Inventory-53 GSI. Additionally, the Brown et al. (2000) sample consisted of slightly younger parents and functioning was assessed with the Symptom Checklist-90-R GSI.

The mean self-reported pediatric health-related quality of life score (M = 70.61, SD = 16.32) as assessed by the Pediatric Quality of Life Inventory (PedsQL; Varni et al., 2001) paralleled those found in another sample of children with SCD (*M* = 65.8, *SD* = 17.3; McClellan et al., 2008). With regards to comparison pediatric populations, the current SCD sample reported QOL scores similar to that of children with asthma (*M* = 74.65, *SD* = 15.79; Varni, Burwinkle,

Rapoff, Kamps, & Olson, 2004), cancer (*M* = 71.90, *SD* = 18.19; Varni, Limbers, & Burwinkle, 2007), obesity (*M* = 67.0, *SD* = 16.3; Schwimmer, Burwinkle, & Varni, 2003), and HIV (*M* = 73.6, *SD* = 13.7; Banerjee, Pensi, & Banerjee, 2010). Moreover, QOL was slightly lower (indicating more negative QOL) than that reported in two healthy samples (*M* = 83.00, *SD* = 14.79; Varni, Seid, & Kurtin, 2001; *M* = 81.08, *SD* = 13.07; Varni, Burwinkle, & Seid, 2006). In light of these findings, health-related quality of life is a significant issue in pediatric SCD. Assessing QOL provides valuable information regarding the functioning of these patients above and beyond biological disease-related factors.

The frequencies of VOC's in children with SCD can vary considerably (Cozzi, Tryon, & Sedlacek, 1987; Gil, Williams, Thompson, & Kinney, 1991) and can be defined as acute versus chronic (Franck et al., 2002). The purpose of the current study was to examine the role of VOC frequency on psychosocial outcomes in caregivers and children. On average, caregivers in the current sample reported that their children experienced approximately 3.55 (*SD* = 4.20) VOC's in the past year. In the literature, retrospective reports of frequency of VOC is a commonly used indicator of disease severity (Brown, Connelly, Rittle, & Clouse, 2006; Schaeffer et al., 1999); genotype, hemoglobin levels, and hospitalizations are other indicators (Barakat, Patterson, Tarazi, and Ely, 2007; Jacob et al., 2003; Lutz, Barakat, Smith-Whitley, Ohene-Frempong, 2004; Zempsky et al., 2008). However, the validity of the aforementioned measures as direct indicators of disease severity remains quite variable (e.g., Barakat, Lash, Lutz, & Nicolau, 2006). Regardless of the difficulty in assessing disease severity, previous literature has consistently found that one's perception of illness severity such as retrospective reports of disease-related occurrences may influence psychosocial adjustment more strongly than objective physiological variables alone (Boekaerts & Roder, 1999; Midence & Elander, 1994; Stuber et al., 1997;

Thompson et al., 1994). Therefore, the frequency of VOC's continues to be an important factor contributing to psychosocial adjustment in families of children with SCD.

Parents of children with SCD reported on the extent to which their child's illness and/or treatment interfere with certain life domains (Devins, 1994; Devins et al., 1993). The mean total parental illness intrusiveness rating (IIRS-P) total score (*M* = 31.42, *SD* = 20.08) for caregivers of children with SCD were similar to that of two samples of parents of children with juvenile rheumatic disease (Andrews et al., 2007; Andrews et al., 2009). With regards to other illness populations, these caregivers reported similar perceived illness intrusiveness than patients diagnosed with angioplasty (*M* = 29.8, *SD* = 14.18), non-insulin dependent diabetes mellitus (*M* = 27.3, *SD* = 14.09), and ulcerative colitis (*M* = 27.6, *SD* = 16.62; for a review see Devins, 2010). However, perceived parental illness intrusiveness in the current sample was lower than that reported in samples of patients diagnosed with fibromyalgia (M = 54.8, *SD* = 17.44), HIV (*M* = 55.2, *SD* = 18.26), end-stage renal disease (*M* = 41.8, *SD* = 15.67), and multiple sclerosis (*M* = 44.8, *SD* = 20.13; for a review see Devins, 2010). It is also likely that some of these differences may be attributable to specific disease-related variables. However, it is important to note that caregivers of children with a chronic illness are often experiencing similar levels of perceived illness intrusiveness as the chronic illness populations themselves. This has been supported in the literature which has found that the general outcome for parents of a child with a chronic illness is similar to that of the child (Timko, Stovel, & Moos, 1992). Thus, it is important to attend to the functioning of parents in conjunction with that of their children since the parenting role involves managing (or supervising their child) medication administration, coping with symptoms of the disease and associated feelings, and often frequent hospital visits and/ or hospitalizations (Moskowitz et al., 2007).

Consistent with hypotheses and previous research, experiencing a greater frequency of VOC's was related to decrements in QOL across domains (physical, emotional, social, and school) in children with SCD. However, this relationship was not mediated by parental perceived illness intrusiveness. Assessing QOL in pediatric populations has been found to be of increasing importance (Drotar, 2004; Matza, Swensen, Flood, Secnic, & Leidy, 2004) as it provides valuable information regarding health and well-being (Pal, 1996). Results of the current study extend previous research that has documented the relation between VOC frequency and QOL in pediatric SCD (Palermo et al., 2002; Panepinto et al., 2005). It might be that the unpredictable nature of increased SCD VOC's taxes children's ability to adequately cope; thus, negatively impacting QOL. Given that these are cross-sectional data, another interpretation is that children with decreased QOL experience a greater frequency of VOC's. However, data with other pediatric SCD samples support the finding that disease-related factors in SCD impact a variety of domains of children's daily of functioning (Dinges et al., 1990; Fuggle et al., 1996; Hurtig et al., 1989; Palermo et al., 2002; Panepinto et al., 2005; Shapiro et al., 1995).

The lack of a mediator relation of VOC's on QOL through IIRS-P may be due to several possibilities. Whereas previous research has documented the relation between IIRS-P and parental indicators of their own adjustment (Andrews et al., 2007; Andrews et al., 2009), this was the first study to examine the relation between parental IIRS-P and child functioning. In addition, previous research has been mixed with regards to the relations among parental stress processing and child psychosocial outcomes (Brown et al., 2000; Casey, Brown, & Bakeman, 2000). It is also possible that parental perceived illness interruptions across domains may simply not be related to child reports of health and well-being or that other variable(s) may drive this relation.

In contrast to previous research (Midence et al., 1993), the frequency of VOC's was not associated with parental psychosocial adjustment (GSI). Moreover, whereas there was a moderate relation between IIRS-P and GSI, IIRS-P did not mediate the VOC-GSI relation. Research with parents of children with JRA found that the relation between IIRS-P and GSI was moderated by child age (Andrews et al., 2009) and racial group membership (i.e., Native American and Causcasian; Andrews et al., 2007). Thus, it is likely that the aforementioned relations are accounted for by a potential fourth variable. Further analyses were conducted to examine the role of child age on the IIRS-P-GSI relation which revealed a significant interaction. Contrary to hypothesis, parents of younger children whom experienced a high frequency of VOC's endorsed relative increases in psychosocial distress mediated by increases in perceived IIRS-P. This relation was not found in caregivers of older children such that the role of developmental inconsistent expectations appeared to not be a factor as hypothesized. Thus, it is possible that older children may be taking on more of the medical management role resulting in less parental distress even in the face of increased perceived illness intrusiveness. Another explanation is that as children get older, parents are better able to adapt to their child's illness and are better able to function. The moderating effect of age was not consistent with that found in the Andrews et al. (2009) sample, which may be due to unique disease-related factors. Specifically, whereas SCD is typically diagnosed via newborn screening, JRA arises during childhood and can be characterized as early- or late-onset (Centers for Disease Control and Prevention, 2010), which would support the adaptation hypothesis in pediatric SCD. Parents of younger children with SCD appeared to be most at risk for psychosocial maladjustment, which can be attributed to the frequency of VOC's mediated by perceived illness intrusiveness. These

findings expand on previous research by specifying a model through which biological disease related variables impact parental perceptions and adjustment in pediatric SCD.

Limitations and Future Directions

In light of interpreting the results of this study, several limitations should be noted. The data collected relied exclusively on self-report. In the absence of corroborating direct observation, ratings from other individuals, or chart-review, it is possible that method variance might have influenced significant relations. Additionally, response bias may have occurred resulting in respondents portraying themselves in a more positive manner. It will be an important future direction to incorporate multiple reporters (e.g., spouse, child, and medical staff), incorporate other forms of measurement such as observational scales or other data collection modalities (i.e., laptop versus paper and pencil measures), and to assess additional indicators of disease severity such as hospitalizations and hemoglobin levels. In addition, data gathered from other sources could be used to support the validity and reliability of the self-report measurement data.

The implementation of the study in an outpatient medical setting (e.g., children's hospital) limits the ability to generalize the results to families presenting for emergent care, which may reflect a population with more severe distress. Additionally, time constraints associated with the site of data collection restrained the quantity and length of measures that could be completed during the typical wait time. Future studies may use more comprehensive measures, which might be completed via the mail, longer appointments, or home visits. Moreover, given the age range of the participants, it will be important to extend these findings to parents of younger children (i.e., less than 8 years old). In tandem, the cross-sectional design of the study limits the ability to make inferences regarding the causal relations between the

independent and dependent variables. Longitudinal or treatment studies would provide richer information about the directionality of VOC's, illness intrusiveness, age, QOL, and adjustment in children and parents of children with SCD.

In addition, cross-sectional research does not provide information about changes over time or the consistency of the data. Though the independent variable of interest, frequency of VOC's, cannot be directly manipulated experimentally it would be important for future researchers to employ longitudinal study designs to account for individual/ developmental changes over time. Specifically, information regarding the impact of the child and parental study variables on adult development /functioning and relationships would be important to understand.

Conclusions

Overall, increases in the frequency of VOC's in pediatric SCD are associated with poorer pediatric QOL and increases in the psychosocial maladjustment of their caregivers. Several demographic variables were found to be associated with pediatric QOL and caregiver psychosocial functioning in the expected directions. Whereas frequency of VOC's was significantly associated with pediatric QOL, parental perceived illness intrusiveness did not mediate this relation. Further, parental perceived illness intrusiveness mediated the relation between frequency of VOC's and parental psychosocial adjustment but only in parents of younger children. In conclusion, this study suggests that the frequency of VOC's is an integral factor in both the well-being of children with SCD and their parents though these findings are contingent upon parental cognitive appraisals and child developmental factors. Findings from this study contain important avenues supporting the development of evidence-based interventions. Specifically, parents perception of how much their child's illness intrudes on their own lives might be an important variable to consider, especially for parents of younger children.

Cognitive-behavioral interventions may be well-suited to ameliorate the effects of disease impact on well-being in both the patient and caregiver.

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Appendices

Appendix A

Figure 1. The Illness Intrusiveness Theoretical Framework

Figure 1. The Illness Intrusiveness Theoretical Framework.

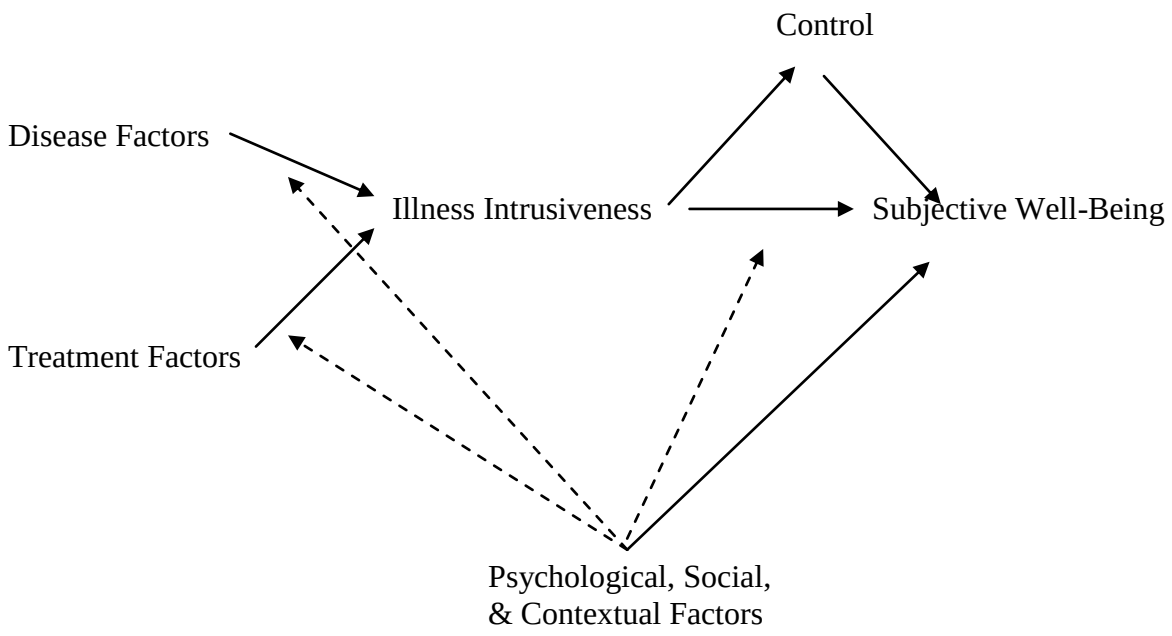

Figure 1. "Disease and treatment factors influence subjective well-being indirectly through their effects on illness intrusiveness. Illness intrusiveness influences subjective well-being directly by reducing gratifying consequences of psychologically meaningful activity and indirectly by reducing personal control. Direct effects are represented by solid arrows. Moderating effects are represented by dashed arrows" (Devins, 1999).

Note. From "Using the illness intrusiveness rating scale to understand health-related quality of life in chronic disease by G.M. Devins, 2009, *Journal of Psychosomatic Research, 68*, p. 591. Copyright 2009 by Elsevier Inc.

Appendix B

Figure 2. Simple Mediation Model 1

Figure 2. Simple mediation model 1.

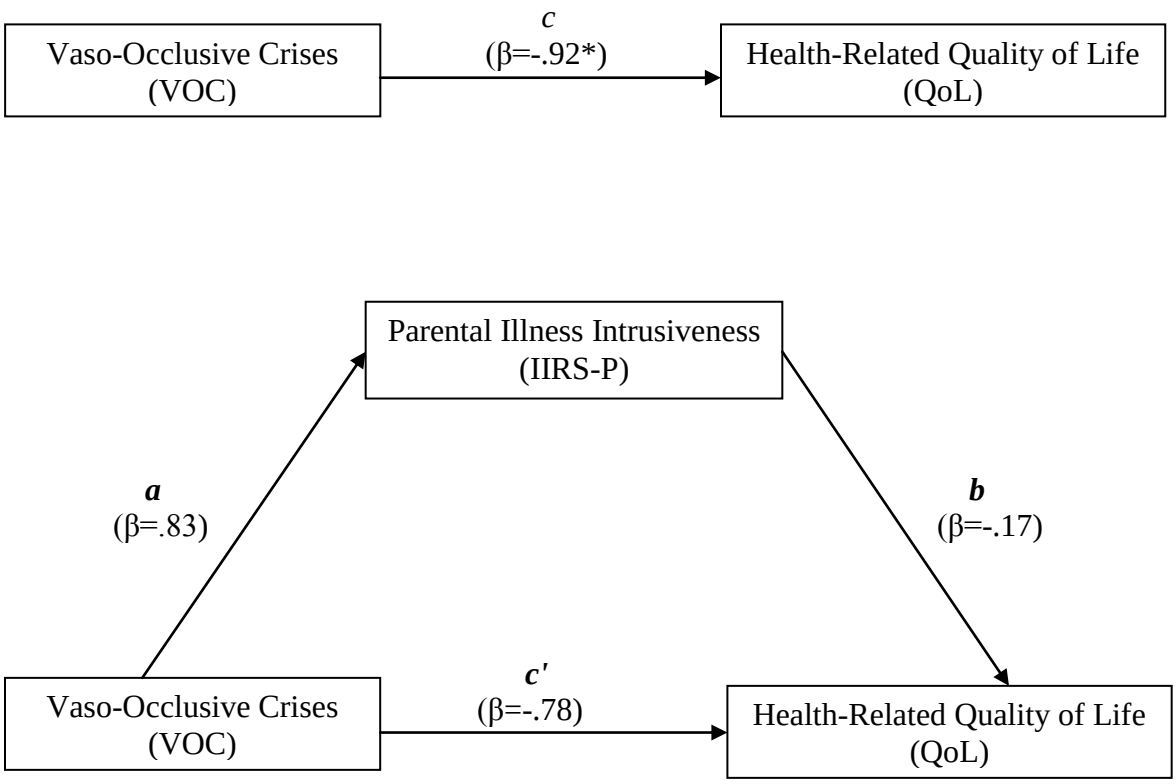

Figure 2. Model 1: Simple mediation model. The indirect relationship of vaso-occlusive crises (VOC) on pediatric health-related quality of life (QoL) through parental illness intrusiveness (IIRS-P), controlling for covariates (i.e., site and comorbid psychiatric diagnoses).

Appendix C

Figure 3. Simple Mediation Model 2

Figure 3. Simple mediation model 2.

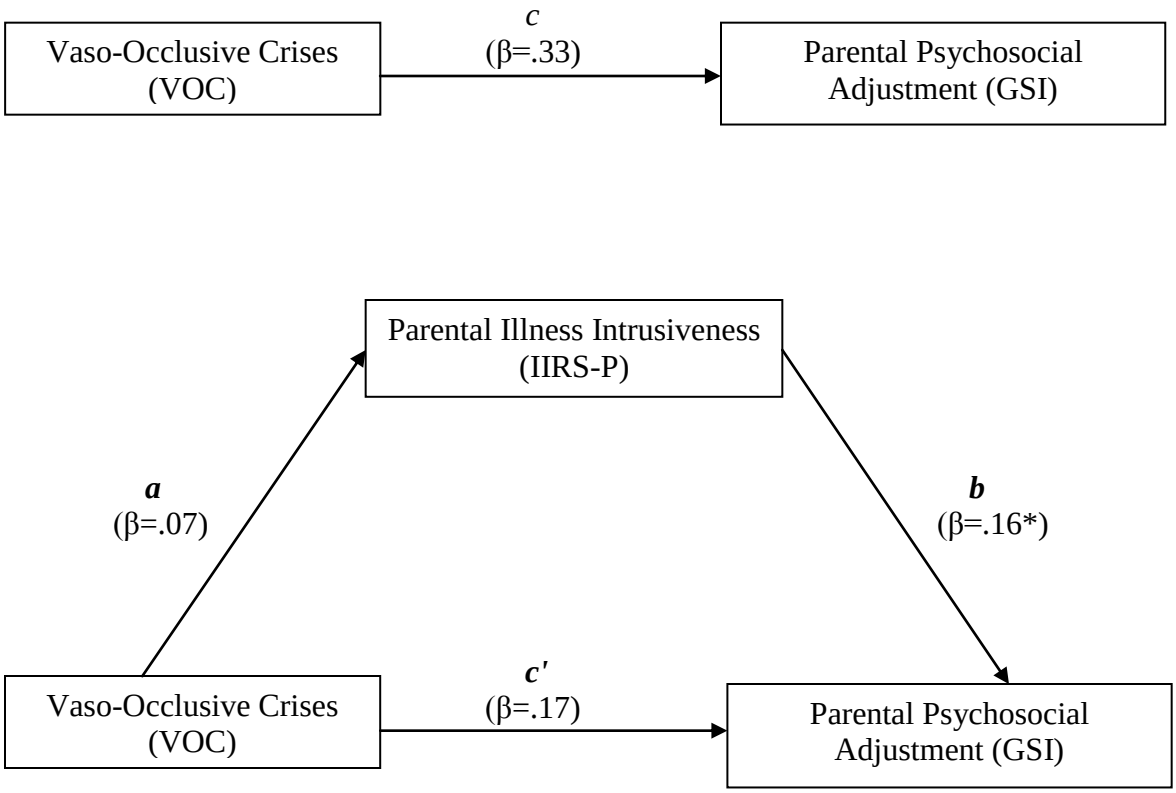

Figure 3. Model 2: Simple mediation model. The indirect relationship of vaso-occlusive crises (VOC) on parental psychosocial adjustment (GSI) through parental illness intrusiveness (IIRS-P), controlling for covariates (i.e., relationship to child, parent age, and family income).

Appendix D

Figure 4. Moderation Model 3

Figure 4. Moderation model 3.

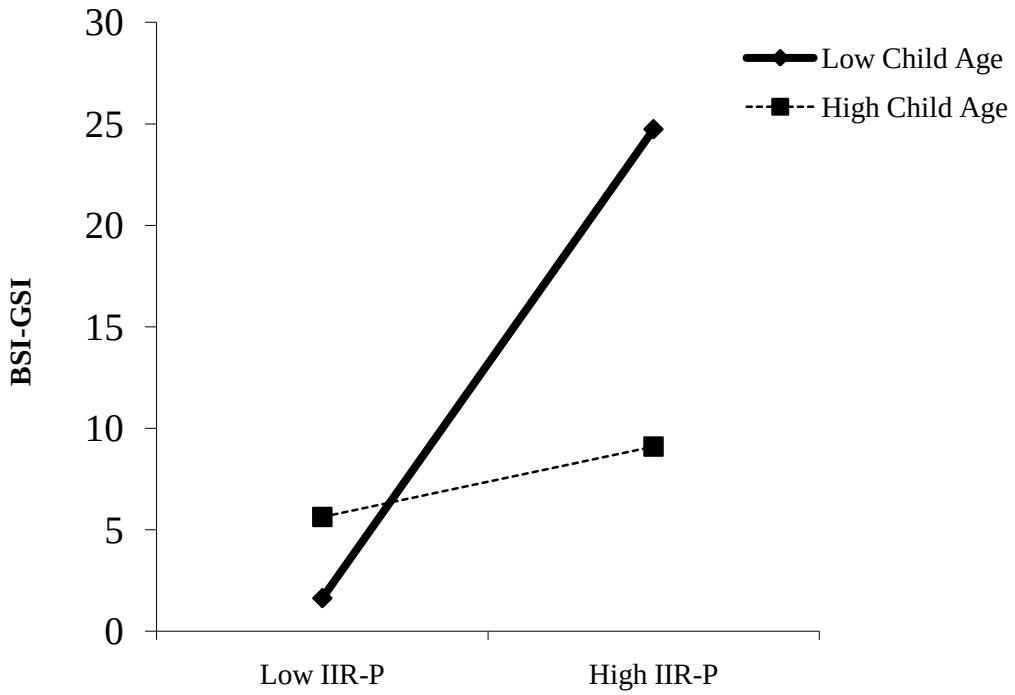

Figure 4. Model 3: The moderating role of child age on the relation between parental illness intrusiveness (IIRS-P) on parental psychosocial adjustment, controlling for covariates (i.e., relation to child, parent age, and family income).

Appendix E

Figure 5. Conditional Mediated Effect

Figure 5. Conditional mediated effect.

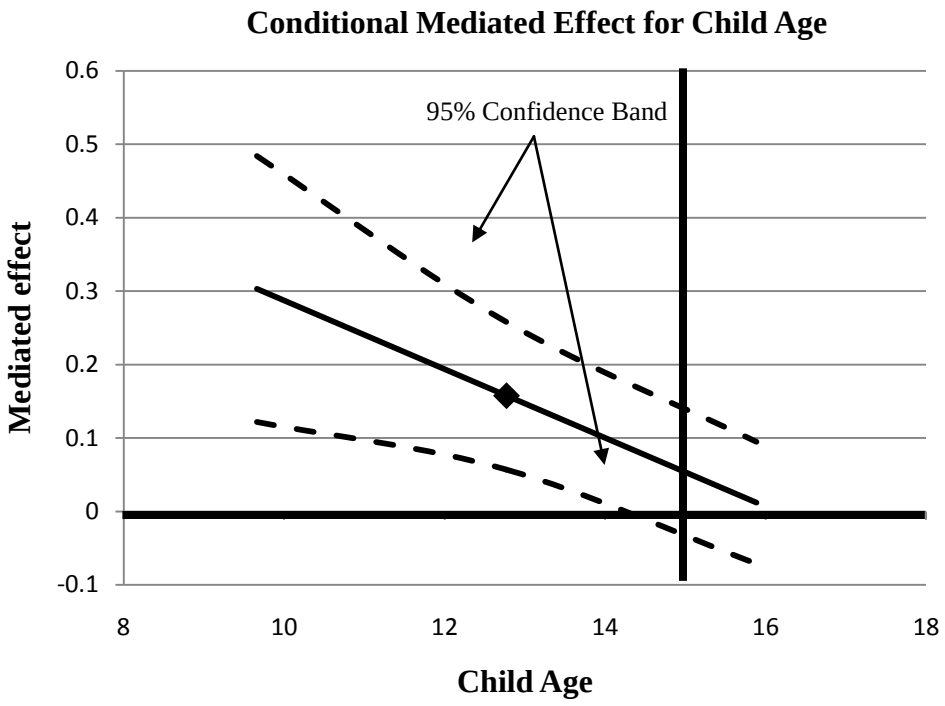

Figure 5. Model 4: Mediated-moderation model. A plot of the indirect effect of vaso-occlusive crises (VOC) on parental psychosocial adjustment (GSI) versus the moderator (child age), with confidence bands. The horizontal line denotes an indirect effect of zero. The vertical line represents the boundary of the region of significance.

Appendix F

Figure 6. Mediated Moderation Model 4

Figure 6. Mediated-moderation model 4.

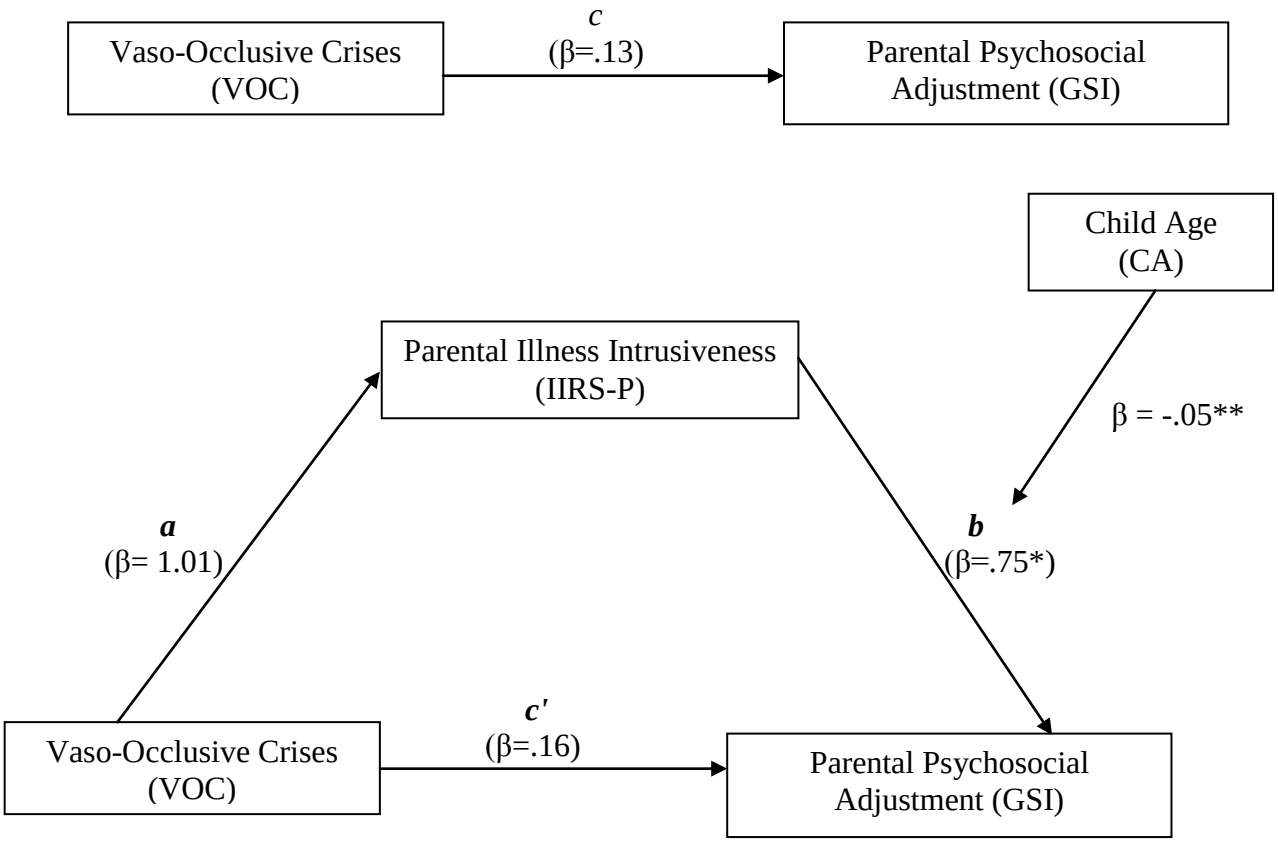

Figure 6. Model 4: Mediated-moderation model. Path diagram of the indirect relationship of vaso-occlusive crises (VOC) on parental psychosocial adjustment (GSI) through parental illness intrusiveness (IIRS-P) moderated by child age, controlling for covariates (i.e., relationship to child, parent age, and family income).

Appendix G

Table 1. Participant Demographics

*Table 1***.** Participant Demographics

Parents

Variable	*M*	*SD*
Age	41.1	8.1
Education Level	13.9	2.1

	N	*Percentile*
Gender		
Female	88	85.4
Male	15	14.6
Relationship to Child		
Mother	86	83.5
Father	9	8.7
Grandparent	6	5.8
Other	2	1.9
Ethnicity		
Not Hispanic/Latino	93	90.3
Hispanic/Latino	1	1.0
Missing	2	1.9
Race		
Black/African American	100	97.1
Native Hawaiian/Pacific Islander	1	1.0
Missing	2	1.9
Marital Status		
Married/Partnered	58	56.3
Single	24	23.3
Divorced	16	15.5
Separated	3	2.9
Missing	2	1.9
Family Income		
Up to $10,000	6	5.8
$10,001 - $20,000	9	8.7
$20,001 - $30,000	12	11.7
$30, 001 - $40,000	14	13.6
$40,001 - $50,000	11	10.7
$50,001 - $60,000	11	10.7
$60,001 - $70,000	5	4.9
$70,001 - $80,000	7	6.8

$80,001 - $90,000	3	2.9
$90,001 and above	16	15.5
Missing	9	8.7

Children

Variable	*M*	*SD*
Age	12.88	3.1
	N	*Percentile*
Gender		
Female	56	54.4
Male	47	45.6
Ethnicity		
Not Hispanic/Latino	88	85.4
Missing	15	14.6
Race		
Black/African American	100	97.1
Asian	1	1.0
Missing	2	1.9
SCD Type		
HbSS	69	67.0
HbSC	13	12.6
Beta Thalessimia	6	5.8
Missing	13	12.6

Appendix H

Table 2. Means, SD, and Intercorrelations

Table 2. Means, SD, and Intercorrelations among Study Variables

Variables	Mean	SD	1.	2.	3.	4.	5.
1. Child Age	12.77	3.11	---	-.05	-.15	.03	.17
2. Parental Illness Intrusiveness	31.42	20.01	---	---	.36**	-.35**	.17
3. Parental Psychosocial Adjustment	6.88	9.24	---	---	---	-.18	.06
4. Health-Related Quality of Life	70.61	16.33	---	---	---	---	-.26*
5. Vaso-Occlusive Crises	3.55	4.20	---	---	---	---	---

Note. *Denotes correlation significant at $p < .05$ (2-tailed). **Denotes correlation significant at $p < .001$ (2 tailed).

Appendix I

Table 3. Simple Mediation Model 1

Table 3. Simple mediation model of vaso-occlusive crises (VOC) on pediatric health-related quality of life (QoL) through parental illness intrusiveness (IIRS-P), controlling for covariates (i.e., site and comorbid psychiatric diagnoses).

Effect	Estimate	Bootstrap SE	t	Significance	95% CI Accelerated and Bias Corrected
Model 1					
c	-.9201	.4146	-2.2192	.0297	
a	.8343	.5484	1.5214	.1326	
b	-.1736	.0880	-1.9737	.0524	
c'	-.7753	.4130	-1.8773	.0646	
a x b	-.1448	.1489	---	---	(LL = -.8529, UL = .1297)

Appendix J

Table 4. Simple Mediation Model 2

Table 4. Simple mediation model of vaso-occlusive crises (VOC) on parental psychosocial adjustment (GSI) through parental illness intrusiveness (IIRS-P), controlling for covariates (i.e., relation to child, parent age, family income).

Effect	Estimate	Bootstrap SE	t	Significance	95% CI Accelerated and Bias Corrected
Model 2					
c	.3322	.2339	1.4200	.1602	
a	1.0119	.5435	1.8619	.0670	
b	.1610	.0491	3.2755	.0017	
c'	.1693	.2242	.7553	.4528	
a x b	.1629	.1837	---	---	(LL = -.0558, UL = .9703)

Appendix K

Table 5. Moderation Model 3

Table 5. Moderation analyses of child age on the relation between parental illness intrusiveness (IIRS-P) on parental psychosocial adjustment, controlling for covariates (i.e., relation to child, parent age, and family income).

Predictor	Beta	SE	t	p
Family Income	-.1244	.3205	-.3883	.6989
Parent Age	-.1886	.1359	-1.3871	.1697
Relation to Child	-1.5890	1.5365	-1.0342	.3046
Child Age	-.0347	.3090	-.1122	.9110
Parental Illness Intrusiveness	.1141	.0270	4.2241	.0001
Parental Illness Intrusiveness X Child Age Interaction	-.0406	.0179	-2.2722	.0261

Appendix L

Table 6. Mediated Moderation Model 4

Table 6. Regression results of the indirect relationship of vaso-occlusive crises (VOC) on parental psychosocial adjustment (GSI) through parental illness intrusiveness (IIRS-P) moderated by child age, controlling for covariates (i.e., relationship to child, parent age, and family income).

Mediator Variable Model

Predictor	*B*	*SE*	*t*	*p*
Constant	-43.9594	11.7489	3.7416	.0004
Vaso-Occlusive Crises	1.0119	.5435	1.8619	.0670

Dependent Variable Model

Predictor	*B*	*SE*	*t*	*p*
Constant	-7.6067	7.6369	-.9960	.3230
Vaso Occlusive Crises	.1284	.2159	.5948	.5541
Parental Illness Intrusiveness	.7455	.2184	3.4137	.0011
Child Age	1.3105	.5796	2.2611	.0272
Parental Illness Intrusiveness X Child Age	-.0462	.0169	-2.7409	.0079

Conditional Effects at Child Age = mean ±1 SD

Child Age	*b*	*SE*	*z*	*LLCI(b)*	*ULCI(b)*
9.6576	.3030	.1810	1.6734	.0253	1.3314
12.7708	.1574	.1055	1.5669	.0117	.6586
15.8840	.0119	.0831	.1432	-.1536	.1718

Appendix M

SCD Background Information Form

SCD Background Information

Questions about the Family

1. Your Relation to Child: ___Mother ___Father ___Grandparent If other, describe: _____

2. Your Gender: ___Male ___Female

3. Your Age: ____

4. Your Ethnicity: ___Hispanic or Latino ___Not Hispanic or Latino

5. Your Race: ___American Indian or Alaska Native ___Asian ___Black or African American ___Native Hawaiian or Other Pacific Islander ___White

6. The highest education level you completed (Please write a number. For example, 8 = completed middle school, 10 = completed sophomore year of high school, 12 = graduated high school, 13 = completed freshman year of college, 16 = graduated college): _____

7. Please describe your occupation: _____

8. Your Marital Status: ___Single ___Married/Partnered ___Separated ___Divorced ___Widowed
   If other, please describe: _____

9. The highest education level your spouse/partner completed (Please write a number. For example, 10 = completed sophomore year of high school, 12 = graduated high school, 13 = completed freshman year of college, 16 = graduated college): ___

10. Please describe your spouse/partner's occupation: _____

11. Please circle your approximate total family income per year:

a. Up to $10,000	f. $50,001 – 60,000
b. $10,001 – 20,000	g. $60,001 – 70,000
c. $20,001 – 30,000	h. $70,001 – 80,000
d. $30,001 – 40,000	i. $80,001 – 90,000

e. $40,001 – 50,000 j. $90,000 and above

12. Do you have a chronic medical condition (e.g., asthma, SCD, diabetes, etc.)? YES NO
    If so, what kind(s) _____

13. Does your spouse/partner have a chronic medical condition? YES NO
    If so, what kind(s) _____

14. Have you been diagnosed with a psychosocial disorder (i.e., anxiety, depression, etc.)? YES NO
    If so, what _____

15. Has your spouse/partner been diagnosed with a psychosocial disorder? YES NO
    If so, what _____

Questions about the Child

16. Child's Gender: ___Male ___Female

17. Child's Date of Birth: ____/____/____

18. Child's Ethnicity: ___Hispanic or Latino ___Not Hispanic or Latino

19. Child's Race: ___American Indian or Alaska Native ___Asian ___Black or African American ___Native Hawaiian or Other Pacific Islander ___White

20. How many *other children* live in the home? ___ What are their ages? _____
    How many children in the home have SCD? _____ How many do not have SCD? _____

21. How many *other adults* live in the home? _____ What are their ages? _____

22. What type of SCD does your child have? _____

23. Does your child have a chronic illness or medical condition besides SCD (e.g., asthma, diabetes)?
    YES NO If so, what? _____

24. Has your child been diagnosed with a psychosocial disorder (i.e., anxiety, depression, etc.)?
YES NO If so, what _____

25. What medication(s) is your child prescribed?
_____

26. Who is responsible for making sure your child takes their medication (i.e., you, child)?
_____

27. When was your child's last SCD related clinic visit? _____

28. When was your child's last SCD related hospitalization? _____

29. How many SCD related pain crises does your child usually experience in one year? _____

30. What major complications has your child experienced related to SCD (i.e., strokes, etc.)?
_____

31. How many days of school has your child missed due to SCD symptoms in the past school year? _____

32. How many days of work have you missed due to your child's SCD symptoms in the past year? _____

33. Would you be willing to allow us to keep you and your child's contact information for follow-up or future research projects? YES NO

If YES, please provide your contact information below:

Your Name: _____

Address: _____

_____

Phone: _____

Appendix N

The Brief Symptom Inventory-18

BSI© 18

DIRECTIONS: Below is a list of problems people sometimes have. Read each one carefully and circle the number that best describes HOW MUCH THAT PROBLEM HAS DISTRESSED OR BOTHERED YOU DURING THE PAST 7 DAYS INCLUDING TODAY. Do not skip any items. If you change your mind, erase your first mark carefully and then fill in your new choice.

HOW MUCH WERE YOU DISTRESSED BY:	NOT AT ALL	A LITTLE BIT	MODERATELY	QUITE A BIT	EXTREMELY
1. Faintness or dizziness	**0**	**1**	**2**	**3**	**4**
2. Feeling no interest in things	**0**	**1**	**2**	**3**	**4**
3. Nervousness or shakiness inside	**0**	**1**	**2**	**3**	**4**
4. Pains in heart or chest	**0**	**1**	**2**	**3**	**4**
5. Feeling lonely	**0**	**1**	**2**	**3**	**4**
6. Feeling tense or keyed up	**0**	**1**	**2**	**3**	**4**
7. Nausea or upset stomach	**0**	**1**	**2**	**3**	**4**
8. Feeling blue	**0**	**1**	**2**	**3**	**4**
9. Suddenly scared for no reason	**0**	**1**	**2**	**3**	**4**
10. Trouble getting your breath	**0**	**1**	**2**	**3**	**4**
11. Feelings of worthlessness	**0**	**1**	**2**	**3**	**4**
12. Spells of terror or panic	**0**	**1**	**2**	**3**	**4**
13. Numbness or tingling in parts of your body	**0**	**1**	**2**	**3**	**4**
14. Feeling hopelessness about the future	**0**	**1**	**2**	**3**	**4**
15. Feeling so restless you couldn’t sit still	**0**	**1**	**2**	**3**	**4**
16. Feeling weak in parts of your body	**0**	**1**	**2**	**3**	**4**
17. Thoughts of ending your life	**0**	**1**	**2**	**3**	**4**
18. Feeling fearful	**0**	**1**	**2**	**3**	**4**

Appendix O

Illness Intrusiveness Rating Scale- Parent Version

Parent Illness Intrusiveness Ratings

Directions: The following items ask about how much your child's illness and/ or its treatment interfere with your life. *Please* ***circle the one number*** *that best describes your current life situation*. Please do not leave any item unanswered.

How much does your child's illness and/ or its treatment interfere with the following:

1. Your feeling of being healthy?

Not At All 1 2 3 4 5 6 7 Very Much

2. The things you eat and drink?

Not At All 1 2 3 4 5 6 7 Very Much

3. Your work (including job, house work, chores, or errands)?

Not At All 1 2 3 4 5 6 7 Very Much

4. Physical recreation or hobbies (such as playing sports, gardening, or other physical hobbies)?

Not At All 1 2 3 4 5 6 7 Very Much

5. Quiet recreation or hobbies (such as reading, TV, music, knitting, etc.)?

Not At All 1 2 3 4 5 6 7 Very Much

6. Your financial situation?

Not At All 1 2 3 4 5 6 7 Very Much

7. Your relationship with your spouse or domestic partner?

Not At All 1 2 3 4 5 6 7 Very Much

8. Your sex life?

Not At All 1 2 3 4 5 6 7 Very Much

9. Your relationship and social activities with your family?

Not At All 1 2 3 4 5 6 7 Very Much

10. Social activities with your friends, neighbors, or groups?

Not At All 1 2 3 4 5 6 7 Very Much

11. Your religious or spiritual activities?

Not At All 1 2 3 4 5 6 7 Very Much

12. Your involvement in community or civic activities?

Not At All 1 2 3 4 5 6 7 Very Much

13. Your self-improvement or self-expression activities?

Not At All 1 2 3 4 5 6 7 Very Much

Appendix P

The Pediatric Quality of Life Inventory

PedsQL™

Pediatric Quality of Life Inventory

Child Report (ages 8-12)

Directions

On the following page is a list of things that might be a problem for you. Please tell us **how much of a problem** each one has been for you during the **past ONE month** by circling:

0 if it is **never** a problem
1 if it is **almost never** a problem
2 if it is **sometimes** a problem
3 if it is **often** a problem
4 if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question please ask for help.

In the past **ONE month**, how much has this been a **problem** for you…

About My Health and Activities (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. It is hard for me to walk more than one block	**0**	**1**	**2**	**3**	**4**
2. It is hard for me to run	**0**	**1**	**2**	**3**	**4**
3. It is hard for me to do sports activity or exercise	**0**	**1**	**2**	**3**	**4**
4. It is hard for me to lift something heavy	**0**	**1**	**2**	**3**	**4**
5. It is hard for me to take a shower or bath by myself	**0**	**1**	**2**	**3**	**4**
6. It is hard for me to do chores around the house	**0**	**1**	**2**	**3**	**4**
7. I hurt or ache	**0**	**1**	**2**	**3**	**4**
8. I have low energy	**0**	**1**	**2**	**3**	**4**

About My Feelings (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. I feel afraid or scared	**0**	**1**	**2**	**3**	**4**
2. I feel sad or blue	**0**	**1**	**2**	**3**	**4**
3. I feel angry	**0**	**1**	**2**	**3**	**4**
4. I have trouble sleeping	**0**	**1**	**2**	**3**	**4**
5. I worry about what will happen to me	**0**	**1**	**2**	**3**	**4**

How I Get Along with Others (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. I have trouble getting along with other kids	**0**	**1**	**2**	**3**	**4**
2. Other kids do not want to be my friend	**0**	**1**	**2**	**3**	**4**
3. Other kids tease me	**0**	**1**	**2**	**3**	**4**
4. I cannot do things that other kids my age can do	**0**	**1**	**2**	**3**	**4**
5. It is hard to keep up when I play with other kids	**0**	**1**	**2**	**3**	**4**

About School (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. It is hard to pay attention in class	**0**	**1**	**2**	**3**	**4**
2. I forget things	**0**	**1**	**2**	**3**	**4**
3. I have trouble keeping up with my schoolwork	**0**	**1**	**2**	**3**	**4**
4. I miss school because of not feeling well	**0**	**1**	**2**	**3**	**4**
5. I miss school to go to the doctor or hospital	**0**	**1**	**2**	**3**	**4**

PedsQL™

Pediatric Quality of Life Inventory

Teen Report (ages 13-18)

Directions

On the following page is a list of things that might be a problem for you. Please tell us **how much of a problem** each one has been for you during the **past ONE month** by circling:

0 if it is **never** a problem
1 if it is **almost never** a problem
2 if it is **sometimes** a problem
3 if it is **often** a problem
4 if it is **almost always** a problem

There are no right or wrong answers.
If you do not understand a question please ask for help.

In the past **ONE month**, how much has this been a **problem** for you…

About My Health and Activities (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. It is hard for me to walk more than one block	**0**	**1**	**2**	**3**	**4**
2. It is hard for me to run	**0**	**1**	**2**	**3**	**4**
3. It is hard for me to do sports activity or exercise	**0**	**1**	**2**	**3**	**4**
4. It is hard for me to lift something heavy	**0**	**1**	**2**	**3**	**4**
5. It is hard for me to take a shower or bath by myself	**0**	**1**	**2**	**3**	**4**
6. It is hard for me to do chores around the house	**0**	**1**	**2**	**3**	**4**
7. I hurt or ache	**0**	**1**	**2**	**3**	**4**
8. I have low energy	**0**	**1**	**2**	**3**	**4**

About My Feelings (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. I feel afraid or scared	**0**	**1**	**2**	**3**	**4**
2. I feel sad or blue	**0**	**1**	**2**	**3**	**4**
3. I feel angry	**0**	**1**	**2**	**3**	**4**
4. I have trouble sleeping	**0**	**1**	**2**	**3**	**4**
5. I worry about what will happen to me	**0**	**1**	**2**	**3**	**4**

How I Get Along with Others (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. I have trouble getting along with other teens	**0**	**1**	**2**	**3**	**4**
2. Other teens do not want to be my friend	**0**	**1**	**2**	**3**	**4**
3. Other teens tease me	**0**	**1**	**2**	**3**	**4**
4. I cannot do things that other teens my age can do	**0**	**1**	**2**	**3**	**4**
5. It is hard to keep up with my peers	**0**	**1**	**2**	**3**	**4**

About School (problems with…)	**Never**	**Almost Never**	**Sometimes**	**Often**	**Almost Always**
1. It is hard to pay attention in class	**0**	**1**	**2**	**3**	**4**
2. I forget things	**0**	**1**	**2**	**3**	**4**
3. I have trouble keeping up with my schoolwork	**0**	**1**	**2**	**3**	**4**
4. I miss school because of not feeling well	**0**	**1**	**2**	**3**	**4**
5. I miss school to go to the doctor or hospital	**0**	**1**	**2**	**3**	**4**