

SICKLE CELL DISEASE AND THE FAMILY:
A PHENOMENOLOGICAL STUDY

by

KEVIN C. GARRETT

B.S., Utah State University, 2003
M.Ed., University of Oregon, 2006

AN ABSTRACT OF A DISSERTATION

submitted in partial fulfillment of the requirements for the degree

DOCTOR OF PHILOSOPHY

School of Family Studies and Human Services
College of Human Ecology

KANSAS STATE UNIVERSITY
Manhattan, Kansas

2014

Abstract

Sickle cell disease (SCD) is a prevalent, pervasive chronic illness. It is a hereditary condition that affects those of African, Mediterranean, Indian, Middle Eastern, and Hispanic/Latino descent. It causes extreme pain for patients and a myriad of other symptoms and complications. The medical issues associated with and the very nature of SCD has the potential to cause psychological distress and related problems for patients. Parents, caregivers, significant others, and family members are similarly affected by a family member with SCD. Applying the Vulnerability-Stress-Adaptation Model, this qualitative study used heterogeneous sampling and explored the experience of three families with SCD. Three main themes emerged from the data, analyzed using thematic analysis: Stress and Challenges, Adapting to and Coping with the Demands of SCD, and Individual and Family Strengths. The pervasiveness and unpredictability of SCD as well as the strengthening effects of having experienced SCD were shared across families, despite their heterogeneity. Clinical implications for families with SCD are discussed.

SICKLE CELL DISEASE AND THE FAMILY:
A PHENOMENOLOGICAL STUDY

by

KEVIN C. GARRETT

B.S., Utah State University, 2003
M.Ed., University of Oregon, 2006

A DISSERTATION

submitted in partial fulfillment of the requirements for the degree

DOCTOR OF PHILOSOPHY

School of Family Studies and Human Services
College of Human Ecology

KANSAS STATE UNIVERSITY
Manhattan, Kansas

2014

Approved by:

Major Professor
Joyce A. Baptist

Copyright

KEVIN C. GARRETT

2014

Abstract

Sickle cell disease (SCD) is a prevalent, pervasive chronic illness. It is a hereditary condition that affects those of African, Mediterranean, Indian, Middle Eastern, and Hispanic/Latino descent. It causes extreme pain for patients and a myriad of other symptoms and complications. The medical issues associated with and the very nature of SCD has the potential to cause psychological distress and related problems for patients. Parents, caregivers, significant others, and family members are similarly affected by a family member with SCD. Applying the Vulnerability-Stress-Adaptation Model, this qualitative study used heterogeneous sampling and explored the experience of three families with SCD. Three main themes emerged from the data, analyzed using thematic analysis: Stress and Challenges, Adapting to and Coping with the Demands of SCD, and Individual and Family Strengths. The pervasiveness and unpredictability of SCD as well as the strengthening effects of having experienced SCD were shared across families, despite their heterogeneity. Clinical implications for families with SCD are discussed.

Table of Contents

Acknowledgements.....	viii
Dedication.....	x
Chapter 1 - Introduction.....	1
Chapter 2 - Theoretical Framework and Literature Review	4
The Vulnerability-Stress-Adaptation Model	4
Sickle Cell Disease (SCD).....	5
Nature of SCD.....	5
Treatment	6
Hereditary Condition	7
Psychosocial and Systemic Issues	8
Pediatric SCD Patients.....	8
Adult SCD Patients.....	9
Parents/Caregivers of SCD Patients.....	10
Patient - Parent/Caregiver Relationships	10
Siblings of SCD Patients.....	11
Families and Chronic Illness.....	11
Purpose of the Study	14
Chapter 3 - Methodology.....	15
Recruitment and Data Collection.....	15
Participants.....	17
The Whitman Family	18
The Jones Family	18
The Putnam Family.....	19
Data Analysis	19
Person of the Researcher.....	21
Chapter 4 - Findings.....	22
Stress and Challenges	22
SCD Symptoms.....	22

Non-SCD Symptoms and Stresses	25
Adapting to and Coping with the Demands of SCD	27
Education and Acceptance	27
The Necessity of Medical Care	29
Family Support and Family Time	32
Extended Family	32
Family Time	33
Individual and Family Strengths	34
Chapter 5 - Discussion	37
Emergent Themes and the VSA Model	37
Contributions to the Empirical Literature	40
Strengths, Limitations and Further Research	41
Clinical Implications	44
References	47
Appendix A - Vulnerability-Stress-Adaptation Model	54
Appendix B - Interview Guide	55
Appendix C - Follow-up Interview Guide	56
Appendix D - Adult Informed Consent Form	57
Appendix E - Parental Consent Form	63
Appendix F - Child Assent Form	68
Appendix G - Rights of Research Subjects Handout	70
Appendix H - Outline of Emergent Themes and Sub-themes	72
Appendix I - Participating Family Demographics Summary	73

Acknowledgements

I have many people to thank for helping me get to where I am today. In fact, I have so many to thank that there is not room enough in this section. Even if their names are not mentioned below, I hope that they know how much they have helped me and that they feel of my sincere appreciation.

As major professor, Dr. Joyce Baptist was always there to encourage, prod, support, and offer suggestions. She has been most helpful and ever-patient. I am especially grateful for her friendship and tutelage.

I would like to thank my other committee members, Drs. Jared Anderson, Karen Myers-Bowman, Valentina Remig and W. David Robinson. Their suggestions, encouragement and time are most appreciated. I especially would like to thank Dr. Robinson for his support and confidence in my abilities.

I would also like to acknowledge all that my wife, Melissa, has done and sacrificed so that I might be in the position that I am in today. Without her love, concern, support, constant prodding, and endless amounts of patience, I would never have completed this dissertation or my degree.

My wonderful children, Colin, Liam, Ainsley, Camden, and Maxwell, are deserving of my appreciation. While I have been absent more than I would like, my children were always excited when I returned home, which lifted my spirits and reminded me of the reason for what I was doing.

I would like to thank my siblings, Chris, Kim, and Wendy. I am very lucky to have such wonderful and supportive siblings. Their encouragement and humor helped me finish this process.

Finally, I would like to thank my parents, Kent and RoseAnn. They instilled in me a love for knowledge and a spirit of creativity. They helped nurture the belief I have that I can accomplish anything worthwhile through confidence in myself, diligence, and hard work.

Dedication

First, this is dedicated to the late Dr. Anthony Jurich, who helped start me out on this journey and who was always encouraging, supportive, and who loved what he did. His passion for research is contagious. Tony, you are greatly missed and you will never be forgotten!

Second, I dedicate this to the research participants who took part in this study. I appreciate your time and sincere interest. Without your participation, I could have never completed this large undertaking. Thank you!

Last, but surely not least, I dedicate this to my sister, Kimberly, who recently passed away after a long battle with cancer. Your humor and candor were refreshing. I hope that I can have even an ounce of the passion that you did for those things in which you believed so strongly. I miss you and love you!

Chapter 1 - Introduction

Sickle Cell Disease (SCD) is a hereditary disease that affects between 90,000 and 100,000 people in the U.S. (Centers for Disease Control [CDC], 2011). SCD is most prevalent among people of African, Mediterranean, Indian, Middle Eastern, and Hispanic/Latino descent (CDC, 2011). Persons with SCD produce sickle cells instead of normal red blood cells (RBCs). Sickle cells are shaped like a sickle, or c-shaped, are stiff, and tend to stick in the cardiovascular system. As opposed to sickle cells, healthy RBCs are biconcave, round, soft, and freely move through the cardiovascular system. When sickle cells stick, they tend to clump together. This clumping often blocks blood flow and results in vaso-occlusive crises (Wagner et al., 2004). These crises generally cause extreme pain for the patient, which frequently result in hospitalizations (Helps, Fuggle, Udwin, & Dick, 2003). SCD, typically diagnosed shortly after birth, is rarely cured (Jenerette, Funk, & Murdaugh, 2005). Instead, it causes physical problems for the patient throughout their lives, especially acute and extreme pain. Understandably, there is a higher prevalence rate of mental health disorders, such as depression, among SCD patients (Helps et al., 2003; Levenson et al., 2008).

SCD is a chronic condition with symptoms that are relapsing and slow to improve (Rolland, 1994a). The longevity of SCD is challenging for families. Because SCD is an illness with a childhood onset, psychotherapeutic interventions need to target entire family systems for extended periods of time. Interventions for adult patients also need to be systemic in nature, taking into account the interconnectedness of families (Rolland, 1994a, 1994b). Because most of the groups affected by SCD are collectivistic in nature, interventions that help all members of the family system are likely to be more effective. For these reasons, it is important that

psychotherapeutic interventions include all pertinent family and group members (Hurtig, Koepke, & Park, 1989).

The current literature does not provide a holistic view of how families are affected by SCD. The paucity of psychosocial studies on SCD-affected families was noted by Brown et al. (1993) and more recently by Thompson et al. (1999). To date, only one qualitative study was found that looks at how the collective family is affected by an adult member with SCD (Strickland, Jackson, Gilead, McGuire, & Quarles, 2001). In this study, adult SCD patients were interviewed together in a focus group, while family members were interviewed together in another focus group. They were not interviewed together. Other qualitative studies have primarily elicited the voice of parents of SCD patients (see Hill, 1994a; Mitchell et al., 2007); only a few studies have included SCD patients (see Strickland et al., 2001).

Also important is that extant studies have included only African American persons or persons who identified as being of African descent. Compared to other ethnic lineages, those of African descent in the U.S. are the most affected by SCD (Strickland et al., 2001). However, SCD also affects persons of Mediterranean, Indian, Middle Eastern, and Hispanic/Latino descent (CDC, 2011). More specifically, Strickland et al. (2001) stated that SCD “primarily affects people of African and Mediterranean descent” (p. 36).

In order to develop family-focused interventions for families with SCD, it is important to understand families’ (including patients’) experiences with SCD, as well as how family members interact with each other in relation to this chronic, often terminal, illness. This study extends the current literature on SCD-affected families by including the voices of SCD patients and their families, who were interviewed together, and by exploring how families interact in relation to SCD. Because SCD is an illness that has to do with the “human experience of individuals and

families” (Rolland, 1994a, p. 129), to understand SCD is to elicit the family’s personal stories that capture the human experience.

Chapter 2 - Theoretical Framework and Literature Review

The Vulnerability-Stress-Adaptation Model

Like any other illness or disability, SCD can lead to either growth or deterioration of relationships (Rolland, 1994b). To understand the process by which a stressor, such as chronic illness, impacts family relationships, the Vulnerability-Stress-Adaptation Model (VSA; Karney & Bradbury, 1995) is applied and viewed through a family systems lens. The VSA provides a practical, overarching framework that offers a comprehensive view of how stressful life events and enduring vulnerabilities have the potential to influence adaptive processes, which in turn have the potential to affect the overall quality of relationships. A figure illustrating the application of the VSA model is provided in Appendix A.

Accordingly, how families adapt when individual characteristics of families and preexisting conditions of individual family members (e.g., vulnerabilities) interact with stresses (e.g., recurrence of illness, loss of a family member) and circumstances (e.g., health status, poverty), impacts the quality of family relationships. Family relational quality can influence family stability – the degree to which relationships in a family stay constant and are resistant to change, and bi-directionally influence adaptive processes.

Enduring vulnerabilities also have the potential to influence, even exacerbate, stressful events, which can in turn influence adaptive processes. In essence, adaptive processes are the ways in which families contend with differences of opinion and familial difficulties and transitions. The choice and implementation of adaptive processes or coping strategies may impact the quality of family relationships.

In keeping with the above extrapolation of the VSA model to family systems (as opposed to marital dyads alone), each family interviewed for this study has a family member who had or currently has SCD. This chronic illness is a circumstance that is stressful for the patient and their families. Each family is comprised of individuals, who each have their own vulnerabilities that have the potential to interact with the stress of managing SCD as a family.

Sickle Cell Disease (SCD)

Nature of SCD

Sickle cell disease (SCD) is the umbrella term used to describe a host of inherited disorders known as hemoglobinopathies, or blood diseases affecting hemoglobin, including sickle cell anemia. SCD refers to disorders characterized by a predominance of hemoglobin S protein in red blood cells (Gold, Treadwell, Weissman, & Vachinsky, 2008). Because of the predominance of hemoglobin S, when red blood cells lose their oxygen, they become sickle-shaped, or c-shaped, and stiff. Sickle cells clump together and block blood flow, which often causes extreme pain for the patient. These pain crises often necessitate immediate hospitalization (Famuyiwa & Akinyanju, 1998; Belgrave & Molock, 1991).

If children experience pain crisis, they are often absent from school for extended periods of time (Wagner et al., 2004; Helps et al., 2003). Shapiro et al. (1995) reported that children, who experience acute pain crises, miss up to an average of six to eight weeks of school per year. As a result of absenteeism, these children may have to repeat grade levels more than once (Nishiura & Whitten, 1980). Similarly, pain crisis in adults with SCD often affects their ability to work, leading to distress in the areas of employment and finances (Morgan & Jackson, 1986).

Another problem associated with SCD is that vaso-occlusive crises, or the blocking of blood flow, can lead to bone underdevelopment and damage (Brown et al., 2000; Schatz,

McClellan, Puffer, Johnson, & Roberts, 2007). Other health problems associated with SCD include organ damage, improper physical development (e.g., stunted growth, elongated limbs), silent or overt neurological damage (e.g., strokes), and cognitive impairment. Children with less severe subtypes of SCD (e.g., those who are at reduced risk of strokes or cognitive impairment) are still about three to four times more likely to incur neurological complications than non-SCD children (Early et al., 1998; Kim, Illes, Kaplan, Reiss, & Atlas, 2002). In addition to the above problems, SCD can lead to increased pneumococcal infections, exacerbation of anemia, delayed puberty, and changes in the spleen (Hurtig, Koepke, & Park, 1989).

Treatment

Currently, there is no universal cure (Sickle Cell Disease Association of America, Southern Connecticut, Inc. [SCDAA], 2010), nor a "definitive cure," for SCD; although bone marrow transplantation and gene therapy appear promising (Mitchell et al., 2007). However, there have been several cases of SCD patients being completely cured of this chronic condition. One of these is discussed in this study.

Although bone marrow transplantation is a promising treatment for SCD, it can be made complicated by the symptoms associated with SCD and, therefore, is often not attempted (Jenerette, Funk, & Murdaugh, 2005). For most patients, SCD symptoms can only be minimized and managed through concerted medical care. The continual treatment for SCD has the potential to increase the life of organs and tissues that can become damaged by reduced blood flow from vaso-occlusive crises. As a result, the life expectancy of those with SCD can be significantly increased (Helps, Fuggle, Udwin, & Dick, 2003; Jenerette, Funk, & Murdaugh, 2005; Platt et al., 1994; Thompson et al., 1999). However, a longer life span is attributed to early screening procedures (Jenerette, Funk, & Murdaugh, 2005).

Due to the genetic disposition of persons of African lineage, making them more susceptible to SCD, screening for SCD is now performed on all African American babies after they are born (CDC, 2008). Early detection of SCD is one method that has increased the lifespan of those affected by SCD by about 20 years. This means that SCD patients now usually live well into their 30s and 40s (Lenoci, Telfair, Cecil, & Edwards, 2002), with some adults living a normal life expectancy (Helps, Fuggle, Udwin, & Dick, 2003).

Medical management for SCD patients includes the use of oral, intramuscular and intravenous pain medication, hydration, blood transfusions, or a combination of all three approaches (Mitchell et al., 2007; Gil, Abrams, Phillips, & Keefe, 1989). There is currently one drug clinically proven to reduce the pain and duration of vaso-occlusive crises in SCD patients -- Hydroxyurea (Mitchell et al., 2007; Jenerette, Funk, & Murdaugh, 2005). Unfortunately, the cost of effective healthcare and medications is costly making it inaccessible to everyone affected by SCD (Burnes, Antle, Williams, & Cook, 2008; Hill, 1994a, 1994b). Therefore, many are left to suffer without any medical assistance. Even those who receive medical care still have to endure pain and possible irreversible damage to their internal organs (Womeodu, Bobo-Mosley, Brown, & Gibson, 1994; Charache, Lubin, & Reid, 1985).

Hereditary Condition

SCD is a hereditary condition that is passed down from generation to generation. If one parent has sickle cell anemia and the other parent is not a carrier, all children will be carriers of the sickle cell trait (SCDAA, 2010). This means that the children will not exhibit any sickle cell syndromes or symptoms (e.g., vaso-occlusive crises, elongated limbs, neurological infarcts) but will still carry the sickle cell trait. If one parent has sickle cell anemia and the other has sickle cell trait, there is a 50% chance that each of their children will have SCD. Two parents with

sickle cell trait have a 25% chance that each child born will have SCD. If both parents have SCD, there is a 100% chance that each of their children will have SCD.

Psychosocial and Systemic Issues

Pediatric SCD Patients

SCD poses significant emotional and mental stressors to patients. Children with SCD are at an increased risk for mental disorders. SCD pediatric patients were found to exhibit depressive symptoms (Brown et al., 1993; Wagner et al., 2004; Wilson Schaeffer et al., 1999), more behavioral problems compared to their non-SCD siblings (Brown et al., 1993), and more developmental and behavioral problems than non-SCD children (Thompson, Armstrong, Kronenberger, Scott, McCabe, Smith et al., 1999). Children with SCD tend to attribute medical complications to something they did or did not do, and are at a slightly elevated risk for psychological distress (Barlow & Ellard, 2006) and decreased overall levels of well-being (Boss & Couden, 2002; Helps et al., 2003).

In addition, Noll et al. (1996) found that SCD plays a role in how childhood peers perceive the patient (e.g., less sociable, less aggressive). Chronic conditions often place physical limitations on the patient causing peers to view them less favorably. Having few friends and feeling ostracized can lead to feelings of loneliness and depression.

Adult SCD Patients

When compared to the psychological literature on pediatric SCD patients, studies on adult SCD patients is limited. However, several studies indicate that, regardless of age, all SCD patients may be at an increased risk for depression (Barrett et al., 1988; Damlouji, Kevesse-Cohen, Charache, Georgopoulos, & Folstein, 1982; Hasan, Hashmi, Alhassen, Lawson, Castro, 2003; Nadel & Protadin, 1977). Additionally, negative affect, negative thinking, passive coping, and somatic awareness were found to be related to several measures of poor health in adult SCD patients (McCrae & Lumley, 1997).

Studies on the management of SCD found increased physical and psychological symptoms, pain severity, and frequency of physician visits among patients who reported lower self-efficacy (Edwards, Telfair, Cecil, & Lenoci, 2000). A longitudinal study of African American SCD patients found that stable and good adjustment were related to lower levels of daily and illness-related stress (Thompson, Gil, Abrams, & Phillips, 1996). These authors also reported that positive adjustment of patients was associated with active coping and efforts to keep emotional states in check. The same was true even for those who engaged in “negative thinking/passive adherence (e.g., catastrophizing, fear self statement, and resting)” (p. 257).

To date, one qualitative study was found in the psychosocial SCD literature that explored how families are collectively affected by an adult member with SCD (Strickland et al., 2001). All participants in this study identified as being African American. Adult SCD patients were grouped together in a focus group and interviewed, while their family members were placed in a separate focus group for their interview. Separate themes emerged from the focus group interview data – themes from adult patients and themes from family members. In this study, themes from the data of adult SCD patients included: (1) Self-care techniques and coping with disabling pain,

(2) Emotional responses of persons with sickle cell disease, (3) Belief by others of drug dependency in persons with sickle cell disease, and (4) Use of religion for coping. Themes that emerged from interview data with family members were the following: (1) Emotional responses of family, (2) The impact of sickle cell disease on family relationships, and (3) Family's concern about others' perceptions of the person with sickle cell disease.

Parents/Caregivers of SCD Patients

Several studies have focused on parents/caregivers of pediatric SCD patients. Evans, Burlew, and Oler (1988) investigated the differences of relationship ratings between one- and two-parent families and their SCD-affected children. They found that two-parent families generally rated their relationships with the SCD-affected child more positively than did one-parent families. Additionally, Famuyiwa and Akinyanju (1998) reported that parents of SCD pediatric patients experience greater amounts of stress than do parents of children with other childhood disorders. However, positive parental/caregiver coping were associated with pediatric SCD patients' coping (Brown et al., 1993). These findings illustrate how stress can be related to the type of illness. Illnesses that are more demanding are more likely to affect the ability of parents/caregivers to adapt and cope (Campbell, 2003; Rolland, 1994a, 1994b).

Patient - Parent/Caregiver Relationships

This relationship is said to work both ways, in that pediatric patients and caregivers both have the potential to influence one another. In other words, coping of the patient and the parent(s)/caregiver(s) is bidirectional, interrelated, and systemic (Campbell, 2003; McDaniel, Hepworth, & Doherty, 1992; Rolland, 1994b). If a child does not cope well with the illness, parents/caregivers can feel guilty (Hill, 1994b) or hopeless when they are not able to help. As a result, these parents/caregivers are at risk of anxiety and depression.

Siblings of SCD Patients

Siblings of patients with SCD are not spared the stress associated with SCD. In their meta-analysis of 51 studies, Sharpe and Rossiter (2002) found that psychological functioning and peer activities, among others, were lower for siblings of SCD pediatric patients than control groups. However, certain positive family characteristics (e.g., high levels of family coping, support and expressiveness, and low levels of conflict) are highly predictive of positive sibling adjustment (Gold et al., 2008). This finding supports the systemic notion of interconnectedness and bidirectionality of the illness, the patient, and the family. The levels of individual and family functioning each have the potential to influence the course of the illness and the patient's well-being (Karney & Bradbury, 1995; McDaniel et al., 1992; Rolland, 1994a).

Families and Chronic Illness

Chronic illnesses each pose unique challenges to patients and families – depending upon the family's developmental stage and its accompanying tasks (Rolland, 1994a). These unique challenges and differences make up Rolland's "Psychosocial Typologies of Illness": (1) onset of the illness, (2) course of the illness, (3) outcome of the illness, (4) type or degree of incapacitation from the illness, and (5) degree of uncertainty surrounding the illness. Within each of these typologies are varying subtypes. For example, within the first typology, *onset of the illness*, there are two subtypes: acute versus gradual. Onset has to do with the ways in which symptoms associated with the condition presented themselves when they first become noticeable (Rolland, 1994a). The type of onset presents different challenges. Gradual onsets are when an illness can be anticipated, while sudden or acute onsets are unexpected (Rolland, 1994a).

SCD has an acute onset. Pain crises from blocked blood flow are often unexpected and can strike at any moment (Helps et al., 2003). These can lead to extended hospital admissions and time away from home, school, or work (Wagner et al., 2004; Helps et al., 2003). Unlike illnesses with a gradual onset, there is no time to prepare physically, emotionally, and/or mentally for the deteriorating nature of an acute onset (Rolland, 1994a).

The second typology, *course of the illness*, presents in three forms -- progressive, constant, and relapsing. Some illnesses can have multiple forms. A *progressive* illness is one in which symptoms are rarely absent; rather, they steadily become worse in a “stepwise fashion” (Rolland, 1994a). On the opposite end of the spectrum the *relapsing* illness where symptoms are experienced for a period of time and then abate for another period. With this subtype, it is only a matter of time before the symptoms flare up again (Rolland, 1994a).

SCD is a primarily relapsing condition. Pain crises come at unexpected times. When symptoms associated with pain crises are experienced, they may last between one to three weeks. Over time, the symptoms lessen and are gone for a time before flaring up again at any moment. SCD also takes on a progressive course. Symptoms can gradually become more severe as they spread – vaso-occlusive crises can block blood flow, which can cause irreversible brain and organ damage, thus progressively worsening the patient’s health and ultimately leading to death.

The third typology, *outcome of the illness*, describes the likelihood of death and the degree to which the illness can shorten one’s life (Rolland, 1994a). Outcome of the illness varies on a continuum from “nonfatal” on one end, to “shortened life span,” or sudden death, in the middle, to “fatal” on the other end. Rolland (1994a) classified sickle cell anemia (one of the SCD types) as shortened life span or sudden death. SCD often leads to a shortened life span. The aggregate effects of the blocking of blood flow is disastrous to the brain, tissues, and organs of

the body (Brown et al., 2000; Schatz et al., 2007). The cumulative effects of vaso-occlusive crises, occurring since childhood, (e.g., mini strokes, splenic changes, bone underdevelopment) often lead SCD patients to only live into their 30s and 40s, even with advancements in medical treatment (Lenoci et al., 2002).

The last typology, *incapacitation*, refers to the impact of the illness on the ability of the patient to function physically and/or mentally. Incapacitation ranges from none to severe. Because SCD is mostly relapsing in course and acute in onset, its degree of incapacitation varies. When the SCD patient is not having a vaso-occlusive crisis and experiences no pain associated with the condition, s/he is not experiencing any degree of incapacitation related to the illness. However, when the patient is experiencing a pain crisis (which is acute and sudden), s/he is likely to experience moderate to severe incapacitation, due to the pain caused by the clumping of sickle cells. As a result, s/he will likely be hospitalized and unable to perform duties at home, work, or school.

Rolland (1994a) excluded *degree of uncertainty* as an official typology of illness. He noted that degree of uncertainty overarches the other typologies explained above and is, therefore, a “metacharacteristic” that influences and “colors” the four typologies: onset, course, outcome and incapacitation of the illness. Uncertainty about any of the above four typologies can create added stresses and challenges for patients and families. Such uncertainty requires “ongoing, strategic problem solving that can exhaust even the most resilient and adaptive families” (Rolland, 1994a, p. 33). When uncertainty is added to an already stressful and trying situation, families often stay in a hypervigilant state. They must be ever-ready to adapt to the challenges and twists and turns thrown at them by the demands of the illness. The psychosocial

strains on families dealing with a life-threatening condition can rival the physical strains on the patient (Rolland, 1994a).

Purpose of the Study

The purpose of this study is to explore the shared experience of families affected by SCD via “thick, rich descriptions” (Patton, 2002) of participants. In other words, it seeks to provide a very personal understanding of the experiences of family units affected by SCD. This study extends the current literature on SCD by including the voice of patients along with other family members. Where the previous qualitative study on adults with SCD sought to better understand this phenomenon but interviewed adult patients and their family members separately (see Strickland et al., 2001), this study interviewed the patient and family members together. This was done so as to provide a more holistic view of participating families’ experiences with SCD.

Additionally, the findings from this study aim to shed light on the divergence and/or convergence of those affected by SCD, but who are in different life and family developmental stages. This is done as the families in this study represent a heterogeneous sample. As is evidenced in the review of literature, there exist only a few contemporary psychosocial SCD studies within the past ten years. This study contributes where research has been lacking. Lastly, this study gives voice to persons of other lineages who are not typically found in the SCD literature, but who are equally affected by SCD.

Given the purposes of this study, the overarching research question guiding this study is: *How are family relational quality and family stability affected by SCD?* Three specific sub-research questions, which seek to answer the larger research question, include the following: (1) *What challenges does SCD pose to families?*, (2) *How do families adapt to and cope with the demands of SCD?* and (3) *What contributes to family relational quality and stability?*

Chapter 3 - Methodology

This study utilized a phenomenological approach to inquiry. This qualitative approach seeks to answer the following question: "What is the meaning, structure, and essence of the lived experience of this phenomenon for this person or group of people?" (Patton, 2002, p. 104). Given the above research questions and purposes of this study, the phenomenological approach is a good fit. Furthermore, this approach is a good fit given its three underlying assumptions: (1) knowledge is socially constructed and, consequently, incomplete; (2) situations can mean a variety of things to a variety of people in a family (Dahl & Boss, 2005); and (3) "language and meaning of everyday life are significant" (Dahl & Boss, 2005, p. 66-68).

Because meaning making and social construction are relative and different for each family member, it was important and "significant" for each family member to have the opportunity to share his/her perspective (Dahl & Boss, 2005). These various perspectives helped paint a broader picture of the whole family's experience (Giorgi, 1985) living with SCD. In this study, the unit of analysis is the family as defined by the patient and participating family members. Families in this study were comprised of (1) the patient with, or who had, SCD and (2) caregiver(s) – parent(s) or spouse.

Recruitment and Data Collection

Multiple methods were used to recruit participants for this study – through a hematologist, and through friends and contacts on Facebook. The second and third families were recruited about two years after the first family completed their interviews. The amount of time between interviews is indicative of the difficulty this researcher had in recruiting for this study. During this two-year interim, three prospective participant families expressed interest in the

study and then backed out before the interview could be conducted, or were never able to be reached again by the researcher after they made initial contact with him.

When prospective participants contacted the researcher, they were given information about the study and the inclusion criteria. Upon agreeing to participate, participants were informed of the incentive to take part in the study, which was in the form of a \$50 gift card to a store of their choice. Two participating families chose to receive the gift card, which was given to them upon completion of the interview. One family chose to give the incentive to a charity of the researcher's choice – a domestic violence shelter for women.

Participants were informed that they were free to withdraw from the study at any time. Their rights as research participants were discussed before the interview began (Appendix G). Informed consent forms also were discussed and signed before the initial interview commenced (see Appendices D, E, and F). The initial interview with the first family was completed in their relative's home and consent forms were discussed in-person. For the second and third families, who resided in different states than the researcher, consent forms were mailed or sent via e-mail to them prior to the interviews. Given the sensitive nature of the things discussed in the interviews, participants were asked if they would like contact information for mental health professionals where they lived. Two of the three families interviewed requested this information and it was provided to them.

Data was collected by the researcher via a face-to-face interview, Skype, and/or phone conferencing. Participants were informed that, after the first interview, a non-mandatory follow-up interview would be conducted. The purpose of the follow-up interview was to allow families the opportunity to share additional information that they may not have thought of during the initial interview and for the researcher to ask follow-up questions. All families who participated

completed the follow-up interview. During the initial interview, the researcher used the Interview Guide (Appendix B) as a template and probed as needed. During the follow-up interviews, participants were initially asked, "Are there things that you thought of since our previous interview that you want to share or elaborate upon?" Other follow-up questions posed to participants are outlined on Appendix C. All interviews were transcribed verbatim by the researcher or a paid transcriptionist prior to data analysis.

Participants

Participating families met two inclusion criteria: (1) have a family member who has or who had SCD and (2) the families were conversant in English. Families were free to decide who constituted their families and who would participate in the study. Participation was voluntary and any family members could decline participation at any time.

The SCD patients in this study varied in age and in their phase and duration of illness. The heterogeneous sampling strategy was utilized in this study. It allows heterogeneity in small samples to be strengths by taking particular interest in any common patterns that emerged from variation (Patton, 2002). This method of sampling provides variation in perspectives, ranging from more typical to more extreme situations. Greater insight into a phenomenon can be gained from viewing the phenomenon from multiple angles. The intent of the analysis is to identify common themes that capture core experiences that cut across participant variation as well as distinctions within these commonalities.

This study includes three families hereon known as the Whitman family, the Jones family, and the Putnam family. The last names of participating families in this study are pseudonyms. The real names of these families are not used so as to protect their confidentiality. Two families identified as African American while one, who is often mistaken as "White,"

identified as being of Mediterranean descent. These families reside in three different regions across the U.S. – the Midwest, the South, and the Eastern Seaboard. A summary of the demographics of each participating family is found in Appendix I.

The Whitman Family

This family is from the Midwest region of the U.S. The SCD patient is a male child who was eight years old at the time of the interview. He was interviewed in-person with his mother. The patient's younger sister, who was three years old at the time of the interview, also had SCD. She was not included in the interview due to her young age and inability to verbalize her experiences. The interview was performed in the patient's maternal grandparents' home. His grandparents chose not to participate in the study. The patient's father was incarcerated at the time of the interview. The patient's mother has a master's degree in a helping profession. At the follow-up interview two weeks after the initial interview, only the patient's mother participated via phone conferencing. The interviews for this family took two hours to complete, when combined.

The Jones Family

The Jones family is from the Southern region of the U.S. The patient in this family was cured of SCD at age seven through a bone marrow transplant. The patient was 20 years old at the time of the interview and lives outside of his parents' home. The patient is currently working but has plans to start college. The patient's father is a doctor. His mother has a master's degree and was working on her second master's degree at the time of the interviews. For the initial interview, the patient was interviewed with both of his parents via Skype using both audio and video systems – making this a face-to-face interview. The patient's teenage sister declined participation in this study. The follow-up interview was performed with the patient and both of

his parents three weeks after the initial interview via phone conference. The interviews for this family took one and one-half hours combined.

The Putnam Family

This family is from the Eastern region of the U.S. The patient is a 35-year old adult female who was diagnosed with SCD approximately one year before the interview. She was raised by adoptive parents since she was an infant. She is of Mediterranean descent and has a graduate degree in a helping profession. The patient and her husband participated in both interviews. The initial interview was performed via Skype. Only the audio feed worked during this interview, so the interviewer was never able to see the patient or her husband. The follow-up interview was conducted four weeks after the initial interview via phone conferencing. The patient has a teenage son who declined to participate in the study. The total time for both of the interviews for this family took a little over two hours.

Data Analysis

Thematic analysis was used to analyze the data (Boyatzis, 1998). This method is used to identify, analyze, and report patterns or themes that merge as being important to the description of the phenomenon under study. Analysis began with the initial reading through of the first transcript. In the second reading through of the transcript, the researcher began coding the data by identifying concepts and their properties, and capturing them in words or phrases that serve as labels for sections of the data. These words/phrases were written in the margins of the interview transcripts. The words/phrases are meaning units about SCD that appeared important to the participating families, and that they wanted the interviewer to better understand. These words/phrases were then transferred onto note cards, where they were compared and contrasted to delineate and extricate relationships between concepts that form the axis of themes derived

from the data (Boyatzis, 1998). Concepts that were related were combined to form sub-themes within larger themes. This process of coding was performed with subsequent transcripts, where themes were compared and contrasted across transcripts. The themes that emerged from this reduction and clustering of concepts formed a comprehensive picture of the participants' collective experience with SCD.

The analysis of data for this study was performed by the researcher with his major professor acting as a verifier. After each transcript was coded by the researcher, the transcript with open codes written in the margins was sent to the major professor. She read through the transcript and verified the coding choice. This process was followed for each transcript. The same process was used with the development of the emergent themes and sub-themes.

The major professor served as verifier on the coding choices and final emergent themes. Additionally, the major professor taking part in this process was utilized to minimize the impact of the researcher's assumptions, experiences, and background on the data. . This was done in an attempt to prevent the researcher's biases from discoloring the true, lived experiences of participating families (Patton, 2002; Creswell, 1998). Furthermore, the researcher explicitly makes his experiences and assumptions known in the "Person of the Researcher" section below to minimize these biases.

The use of member checking (Oktay, Jacobson, & Fisher, 2013; Patton, 2002) was attempted to improve the credibility and dependability of this study (Creswell, 1998; Merriam, 1998). Participants were given the opportunity to review the final themes that emerged from their interview data. Two of the three families were provided with the emergent themes, per their requests. However, none of the participating families chose to provide additional feedback after receiving these themes.

Person of the Researcher

I am a White male who was raised in a well-served community. My experiences and knowledge of ethnic minority and underserved communities is limited. Despite having been raised in a predominantly privileged and well-protected environment, I have been told about atrocities experienced by those that I serve in my clinical work as a therapist. As such, I have compassion and a reference point for persons who have had to experience hardships that I have not had to endure. In addition, I have witnessed many of my clients transcending life's atrocities. In other words, I have seen them lead productive lives, in spite of their past traumas and hardships. I believe that with the appropriate resources (i.e., financial assistance or security) and support, it is possible to not only survive but to use these experiences to build resiliency. According to the old adage: "What doesn't kill you, only makes you stronger."

It is my assumption that the majority of families (and individual family members) interviewed for this study will generally find that their family relationships and stability have been positively affected (e.g., strengthened) by SCD. With this assumption in mind, it is paramount that I do not impose this belief on participants and, eventually, on the meaning units and ultimately the "structure of the experience" (Dahl & Boss, 2002, p. 74) of SCD-affected families. I will need to depend on the participants' illustration of their lived experience to fully understand their lives. Any assumptions that I have of my participants might not be accurate as it is from the standpoint of a privileged male. For example, an assumption that I tend to have is that families are similarly resilient, which might not be true as not all families are resilient.

Chapter 4 - Findings

Despite the heterogeneity of the age when the diagnosis was given, the progression of the illness, and economic level of these families, common shared experiences across families emerged from the data. These commonalities or convergent experiences across the families are captured by the following themes: *Stress and Challenges*, *Adapting to and Coping with the Demands of SCD*, and *Individual and Family Strengths*. These themes respectively provide answers to each research question guiding this study. Each theme is explained and elaborated upon below with its accompanying sub-themes. An outline of the emergent themes and sub-themes are found in Appendix H.

Stress and Challenges

This theme relates to specific hardships associated with the SCD diagnosis that participating families have endured. This theme answers the first research question: *What challenges does SCD pose to families?* Some hardships were specific to SCD and some were not. Sub-themes include: (1) *SCD Symptoms*, and (2) *Non-SCD Symptoms and Stresses*.

SCD Symptoms

All patients in participating families have experienced SCD specific symptoms, such as pain crises that required hospitalization, and had undergone at least one form of medical procedure or surgery in their lifetime. The patient in the Whitman family has had several pain crises that required medical attention. The patient in the Putnam family has lived most of her life in pain before finally being diagnosed with SCD one year ago. She still experiences pain crises and knows that she will have more surgeries in the future: “With SCD, everything starts to die.”

The patient in the Jones family, before being cured of SCD, experienced pain crises and had his spleen removed. Another stressor associated with SCD relates to the literature on pediatric SCD patients. For example, one study illustrated how non-SCD peers often perceive the patient less favorably (Noll et al., 1996). Such was the case with the patient in the Jones family. His father recalled how a stroke changed the patient and how he was perceived differently by peers following this:

I think one of the things that sticks out in my mind was after [the patient] had a stroke, [the patient] was in Montessori school and, to be honest with you, he was kind of a holy terror before he had the stroke...I remember [he] held down this kid and he bit him. But anyway...after he had the stroke, he was in the hospital for a month, so he wasn't in the Montessori school for about a month...he had done a little bit of rehab, sorta, kinda...but he wasn't quite, um, rehabilitated. He wasn't quite as stable on his feet as he used to be, and he was chasing the other kids around like he used to, and one of the little girls asked the teacher why he didn't run as well as he used to, and, you know, what was wrong with him. But I remember that, and that was one of the things that changed immediately after his stroke.

The patient in the “J” family, shared some memories of stressors that were spurred by the things his parents were recalling during the interview. He shared, “I remember, um, now that, when I heard my parents were talking about it. I do remember...some of the symptoms. I do remember the pain episodes...I do remember swelling...” He later recalled being in the hospital “for a long time” following his bone marrow transplant.

The patient's mother in the Whitman family reported that her daughter, who was three at the time of the interview, experienced worse SCD symptoms than her son. While only three

years old, her daughter has already had her spleen, appendix, and gall bladder removed because of SCD.

As the patient in the Putnam family was not of African lineage, she was not screened for SCD at an early age. Moreover, she was adopted at a very young age and appears "White." She said that one doctor, when it was discovered that she had SCD, exclaimed to her, "But you're White!" This patient expressed several times in her interview with this researcher that she took part in this study to bring "awareness." She wished to bring awareness about SCD affecting those of other lineages, even those who appear "White" and are not typically diagnosed with this chronic illness. Similar to other SCD patients, her stresses revolve around excruciating pain episodes, anemia, an elevated white blood cell count, and having her gall bladder removed. In addition, she was given the "run around" by medical providers before she was finally diagnosed with SCD one year ago. The patient shared:

They said I had a white blood cell count of a leukemia patient... Yeah, they thought I had lymphoma, they thought I had all these things, but they couldn't find any cancer. They passed me from doctor to doctor, they sent me to a heart specialist because of the wooziness and dizziness, and almost passing out, and they'd see my blood pressure really go low and they were saying, "Well, we can see with your cells that you're a bit anemic." They never really checked. They would say, "Well we're going to put you on this iron diet," and then they started putting me on...like a junk food diet and I was a health nut.

In the "P" family, a big stressor is the patient's son's response to his mother's SCD diagnosis. The patient reported that her son is still coming to terms with her diagnosis, especially the likelihood of her shortened lifespan. The patient shared:

My son decided he didn't want to participate. I think he just, I just think this is still too sensitive for him... Since he just found out about this last year... My main concern has been my son because it's just, to be honest, I mean, it's kind of a blow to him. He's 15. He found out when he was 14...

Non-SCD Symptoms and Stresses

While being treated for symptoms or complications associated with SCD, or before being diagnosed with SCD – as in the case of the Putnam family, patients and families experienced stresses related to other medical issues. For example, the patient's mother in the Whitman family reported that she has to worry more about the asthma-related symptoms and/or complications for the patient:

I know I need to watch him more because he has mild asthma, so with him, I really have to stay on top of him to take his inhaler and watch his breathing... I think really with him when he gets, when the weather changes and he's being very active he gets to coughing. And so, when he gets coughing, that's when I give him his treatment and that's when it will stop... And that's how I've learned to watch his breathing thing.

The adult patient in the Putnam family was found to have diverticulosis about the same time she had her gallbladder removed. These were both diagnosed and treated before she was diagnosed with SCD. The patient in the Jones family, as aforementioned, was hospitalized for "multiple infections" regarding his sinuses and ears. These were not necessarily related to or caused by his SCD. Patients and families experienced stresses that were not health-related. In the Whitman family, one of the biggest stresses was the patient's father's incarceration. The mother shared several times in the interview how difficult this was for her. She did not have the support of her husband (the patient's father) during difficult times. She reported:

But that was hard because, well, the sickle cell and dealing with that, um, it's hard on [the patient and his sister] but it's also hard on me because a lot of times, even though I have my family, my parents, it's me that has to deal with all the news and deal with all the decisions and, you know, [patient's sister] had her surgery [where her gallbladder, spleen, and appendix were removed], I had my family with me, but, at the end of the day, it was just me in the hospital room with her. And it was me taking her home, and me at home with her when she was recuperating.

The mother in the Jones family shared that it was stressful when her husband (the patient's father) was deployed. While he was deployed, she recalled that the patient was back in the hospital with another infection:

[Patient's father; her husband] was deployed, I guess. And I think it was right, shortly after we moved to [state], and [the patient] wound up being in the hospital for some kind of infection. And, all I can remember...about that stay is that I was trying to go to sleep and [patient] would not let me sleep. He kept telling me to walk, so I'm pulling him on the floor in this wagon, I mean, I'm just like walking around and around and around in the hospital pulling him in this wagon...And I think I went home for somethin' and a friend of, of someone who worked with [patient's father], a friend of ours, came in and they had awakened [patient], and so he's like freaking out because I wasn't there.

For the Putnam family, a large external stressor was caused by the patient's brother when they were living in the same house with him. They reported that her brother stole money from them and stole the patient's pain medication on five different occasions. The patient's husband reported that he did stand up to his wife's brother about these behaviors. However, her husband reported that he had to keep his anger in check, as he was grateful that the patient's father was

helping them by giving them a place to live that was rent-free, and was also helping them with medical expenses. He reported that there was a good chance they would have been kicked out of the patient's father's home if he were to have physically fought with the patient's brother who was favored by the patient's father.

Adapting to and Coping with the Demands of SCD

Families had to learn about SCD and the demands of the illness. This required the acquisition of new skills that took time and patience. Each family went through, or is going through, a stage in which they had to be constantly on the lookout for SCD's symptoms and progression. The vigilance required and the level of proactive response to the illness has become, or was, a way of life for these families. This theme answers the second research question: *How do families adapt to and cope with the demands of SCD?* Three sub-themes emerged: (1) *Education and Acceptance*, (2) *the Necessity of Medical Care*, and (3) *Family Support and Family Time*. These illustrate the coping strategies and processes of adaptation that helped SCD-affected families.

Education and Acceptance

Following the diagnosis of the patient, there was a process of adaptation and learning for the families. Individuals within each family system had their own emotions and reactions to the diagnosis, which changed over time. Where the SCD diagnosis was given more recently, family members are still actively engaged in the process of coming to terms with the diagnosis and accepting its accompanying stressors and prognosis.

The mother of the Whitman family, whose son and daughter were both diagnosed with SCD, described how she feared losing her son when she first learned of his diagnosis. She described her reaction:

I was crying a lot. I called up a friend and I spoke with her and kind of told her what was going on. I didn't – honestly at the time, when I first heard that it – fatal disease was the first thing that came to my mind because growing up I used to hear about people with sickle cell and they always died young or had all these problems, so, um, that's what I was most distraught about...

However, having a son with SCD helped this mother better handle the news of her second child's SCD diagnosis:

I did not think [daughter] was going to have [SCD] because, when I was pregnant with [the patient], I was very anemic and when I was pregnant with [daughter], I wasn't. So I just assumed that she wasn't going to have it. So, when they called to tell me that, that was just another big shocker. But I handled it a little bit better because by the time I had [daughter], [the patient] was about five, he hadn't had a transfusion, um, and he wasn't getting, um, he wasn't the typical sickle cell patient. So I expected her to be the same way.

She attributed her ability to accept her daughter's diagnosis with less sadness and fear to the research and learning about SCD that she conducted after her son was diagnosed. She shared, "But the more I started doing research...I guess the older he got, the more I realized that it was not as bad as it had been and not as bad as it could be."

The mother in the Jones family had a similar reaction to her son's SCD diagnosis. She recalled that she was home alone when she learned of the news: "I was pretty upset. Because I didn't, that wasn't what I was expecting...I was crying." She shared several things that helped her along the process of adapting to her son's diagnosis. One of these was receiving support from her husband (the patient's father).

The mother in the Jones family shared that she learned a lot about SCD from her husband, who is a doctor; from the patient's pediatrician and hematologist; as well as from books, and social workers in the hospital. The parents shared that they learned that SCD can be cured through bone marrow transplants while attending a SCD conference. This one piece of information was ultimately extremely helpful, as the patient in this family was completely cured of SCD at the age of seven through bone marrow transplantation. The Jones family also learned about pre-implantation testing from a Public Broadcasting Service television special. This procedure was done before the patient's younger sister was conceived, thus eliminating the chances of the sister having SCD.

The patient and her husband in the Putnam family said that they were still learning to adapt to the patient's diagnosis, which was given only one year ago. The patient said, "We've had to adapt." Her husband quipped, "If you don't adapt, you die." The patient shared that she knows what the illness would eventually require of her and that she needed to accept the reality of it in order to be there for her family:

I've had times where I've broken down...knowing that I can't be as much as I want to for my family, you know, I can't do everything I want to do and knowing that I may not always be here. I'm already having problems with my kidneys...I know dialysis will happen eventually and just trying to accept some of these realities and having to sometimes say, "I can't do this, I hate it!" because I want to be able to do everything for my family that someone normally would be able to.

The Necessity of Medical Care

Families shared that, over time, they learned when professional medical care for the patient was necessary. This process involved others in the family becoming more attuned to the

symptoms of the patient, and learning which symptoms were indicative of something urgent requiring immediate medical attention. The severity of symptoms and the cost of medical care often influenced whether families sought medical treatment. The lack of financial resources also resulted in another form of adaptation -- one patient learning to increase her pain tolerance.

In the Putnam family, there were times when the patient would not go to the doctor or hospital because she and her husband were paying for her medical care completely on their own. They shared that they had accrued many medical bills and debt as a result. Due to this, as well as the patient stating that, before she was diagnosed with SCD, others thought she was "faking" her pain, she "built up a high pain tolerance." She would frequently not receive medical care as a result. In other words, she learned to adapt to high levels of pain without seeking medical treatment, or she learned to care for most of her medical issues on her own. However, due to this high pain tolerance, she did not always know when medical care was necessary. Her husband recalled one occasion when the patient's nose was bleeding "profusely" and would not stop. He said that he had to almost literally "shove" the patient in the car to get her to go the hospital, as she was refusing to go due to the expense. He said that it was good that she went to the hospital, as medical providers had to eventually use "cocaine" to stop the bleeding because normal cauterization procedures had not worked. For the patient and her husband in the Putnam family, they are still learning when the patient needs to seek medical attention. They shared that they now have health insurance, and seeking the necessary medical care will not be such an issue, as they will no longer have to pay as many out-of-pocket expenses.

Another reported factor related to this sub-theme dealt with the family trying to help the patient have a "normal" existence. The patient's mother in the Whitman family admitted to being overprotective of and "hovering" over the patient when he was younger. She used to worry

extensively about him. She frequently took him to the doctor or hospital “for every little thing.” She was not sure earlier in his life if he was experiencing “typical boy stuff” or symptoms related to his SCD. However, as the patient and his sister have gotten older, their mother has learned when to seek medical care. She said:

I...am less fearful of somebody getting a fever. I don't immediately call the doctor or rush to the hospital. If [the patient] complains about his stomach hurting or his leg or something hurting, um, I learned to ask him probing questions. I know where to check him at and...I'm, um, learning when to be worried and when not to be worried. So that's changed a lot.

The Jones family learned early on when to take the patient to the hospital and when not to, so as to try to help him have a "normal" existence. The patient's parents said that they usually treated his SCD pain symptoms at home with hot pads and over-the-counter pain medication. On one or two occasions, the patient was treated at home with narcotics, as his father was a doctor. The patient's parents recalled only taking the patient to the hospital on one occasion for a SCD-related pain crisis. However, they recalled that they had to take him to the hospital for “multiple infections” and “running a temperature” on numerous occasions. They reported that, before he had his bone marrow transplant, the patient had “a splenectomy” and “multiple infections” – specifically, “multiple sinus” and “multiple ear infections.” After his splenectomy, the patient had to have blood transfusions. The parents tried spacing out the transfusions so that the patient would miss as little school as possible. When they could help him at home with medical issues, they would. This was their way of trying to help the patient have “a normal existence.”

Family Support and Family Time

Families managed SCD related stresses through the support of their extended family. They also coped with illness-related stresses through family activities and spending time together. Family activities and extended family support helped provide normalcy and hope.

Extended Family

Patients and families noted that their extended families were a big help by providing emotional and physical support, and financial assistance. The mother in the Whitman family emphasized her reliance on her family for support and assistance: “I think I’ve coped really well. You know, my parents...are wonderful. They help me through everything!” To illustrate, she said that she developed a plan with her extended family, in case one of her children has to go to the hospital suddenly. If this occurred, her extended family watched the other child so she could take the acutely ill child to the hospital. She continued to say, “It takes a village to raise a child,” and provided an exemplar where her mother came to stay with her shortly after the patient was diagnosed with SCD as an infant. Her sister (the patient’s aunt) also helped by taking the patient and his younger sister to activities. At the time of the interview, the mother in the “W” family was a single parent because her husband was incarcerated. During this time, her father served as a "father-figure" to the patient, and did “guy things” with the patient, while the patient's maternal grandmother offered emotional support.

The Jones family shared numerous ways in which the patient’s maternal extended family offered support. The patient’s maternal grandmother and great-grandmother frequently came to stay with the family. The patient’s mother reported that her mother stayed with them off and on for 18 months, and that her grandmother (the patient’s great-grandmother) stayed with them for almost four years so the patient's mother could remain employed after they first learned of the

patient's diagnosis. This family attributed their ability to cope to having a "strong support system."

For the Putnam family, the support from extended family (the patient's father) included financial assistance, although the patient described her relationship with her father as strained. Her father helped pay for medical expenses "so we didn't accrue more debt," and allowed this family to live rent-free with him.

Family Time

Having intentional family time together was identified as important for all families, as it helped them cope with the patient's diagnosis and accompanying stresses. The patient from the Putnam family reported that, shortly after she was diagnosed with SCD, they took a family trip to Walt Disney World. She said, "We really couldn't afford it, but we needed some family time together, you know, we just found out I had a terminal disease and everything." She continued, "Anyway, we went to Disney World for a week just to have some family time." The patient's husband in the Putnam family added, "Well, I continually look at it...she's still got plenty of years left in her. I can enjoy 'em or I can mope about and not enjoy them. I think I'd prefer enjoying my time with my wife."

Although the Jones family is in a different stage of life than the Whitman family, both families shared that doing things together, including taking trips, helped build closeness. In the Jones family, where the patient was cured of SCD at the age of seven, activities and family trips served as ways that they coped with the patient's SCD when he was younger. As the mother reported, "I've spent more time, I think we've both spent a lot of time with the kids. But now that they're older, we don't do, like everybody has their own things that they like to do."

For the Whitman family, observing and knowing that her son could engage in activities with non-SCD children helped her cope. She said, “I’ve coped well just knowing that he can do a lot of more normal things that other children can do.” For example, she and her son discussed how he will be playing tackle football in the near future.

Individual and Family Strengths

The collective strength of participating families was made up of the individual strengths and contributions of each family member. Also, the relationships within families allowed families to remain strong as they helped each other cope with the accompanying challenges of SCD. The support, care and concern that family members had for each other were evident in the relationships they had fostered. Also, this care and concern were evident in the compliments family members paid each other. This third theme answers the third research question: *What contributes to family relational quality and stability?*

In the Putnam family, the patient reported that her husband is supportive, in that he took part in this study because he knew it was important to her. She also shared that her husband is willing to attend therapy with her because of how important it is to her, though he would prefer not to attend therapy. The patient’s husband shared many positive traits that he appreciates about her. He said, “She’s tougher than she lets on,” and “She has a lot of willpower going for her.” He also reported that she is a “good cook,” “multi-talented,” “complex,” and “a fast, deep-thinker.” The patient reported that she is “stubborn” but is often “very determined.” Her husband reframed her comment about being stubborn and said that “she has a lot of perseverance.” Within the context of the patient battling with SCD, being “determined” or “stubborn” were presented as strengths, in that the patient was not giving up or giving in to SCD, despite its accompanying stresses.

The patient in the Putnam family commented that her husband “is very good at being able to come into an emotional situation and being able to handle it.” Additionally, she said that her father called her husband “the diplomat of the family.” Regarding how supportive her husband is, the patient stated that her husband provided her with the energy and support to seek multiple doctors and to take part in numerous medical tests, as he did not believe that she was “faking” her pain or symptoms. She said that it was because of his relentless efforts that doctors finally discovered that she had SCD. The husband shared:

I knew she wasn't faking it 'cause I actually have an allergy to seafood, that's one of those things that people...people with allergies, a lot of times get chalked up to "Hey, he's faking it." I can empathize in that situation because I've been through it a little bit, so that made me give credit to her so that I...there's some more going on here...It didn't fit, so I kept pushing her to get more answers. So, part of the [medical] debt is mostly my fault, but that's okay because I kept pushing. But I'd rather have answers than not have answers. Again, with knowing what you have it becomes easier for you to be able to assist the doctors in taking care of yourself.

Ultimately, both the patient and husband in the Putnam family agreed that the patient's SCD has “Benefited us a little bit in the strengthening of our relationship.” They admitted that coping with SCD has helped them build a stronger relationship and a closer family. This is true even with the patient only being diagnosed with SCD one year previously.

The patient's mother in the Whitman family reported, “We're a close family.” She agreed that her family is tight-knit, and reported similar things to being a close family several times throughout the interviews. Additionally, she reported that the patient, as he has gotten older, has done a nice job of taking care of his own health and medical needs. She said that he keeps

himself hydrated, rests when he needs to, and often administers his own asthma medication and treatments by himself. The patient shared some very positive things about his mother, especially related to how much she cares and often worries about him and his sister, though he wished that his mother would not worry so much. Through all that the patient, his younger sister, and mother have been through related to SCD, the mother reported that she has learned “We can get through it!”

In the Jones family, the parents supported one another and helped the other cope. The patient’s father reported that he was “shocked” at first after finding out about his son’s diagnosis. However, he said that his primary goal was to keep his wife “calm” during this stressful time. The patient’s mother said that her husband will usually calm her down when she gets upset. She stated that she will initially, at times, be very upset; will calm herself; and then will know what she needs to do to work through the stressful situation. In reference to how the patient’s father and mother support each other and made a good team, the father said, “There is a calm, pervasive unshakeableness that we have together.”

The researcher observed the Jones family’s level of support for their child during the interviews. At one point in the interview, when the patient started sharing something but then said it was not important, his parents encouraged him to say what he was originally going to share. They said, “Be your own advocate!” and they both encouraged him. He ended up sharing his thoughts in the interview, which were related to something he recalled when he was younger. Additionally in describing their family, the patient’s father shared, “Our family is resilient!”

Chapter 5 - Discussion

The themes above speak to the pervasiveness and unpredictability of SCD, and family strength. The pervasiveness and unpredictability of SCD that infects every level of the family system is well supported by the literature on chronic illness and families (e.g., Campbell, 2003; McDaniel et al., 1992; Rolland, 1994a, 1994b). The pervasive and unpredictable nature of SCD is now explored further as it relates to the VSA Model (Karney & Bradbury, 1995).

Emergent Themes and the VSA Model

SCD has an infectious nature that cannot be contained – it permeates whole family systems (immediate and extended), as well as other related systems – professional life (for adults) and school (for children) (Anie, 2005; Helps et al., 2003; Nishiura & Whitten, 1980; Rolland, 1994a; Strickland et al., 2001; Wagner et al., 2004). The pervasive nature of SCD means that families have to be vigilant and selective when it comes to involvement in activities that non-SCD affected families can take for granted. SCD oftentimes becomes the focus of parents or spouses who have to be attuned to its symptoms. This stress is exacerbated if the patient has other medical conditions that make him/her more vulnerable (Karney & Bradbury, 1995). Vulnerability to pain and other medical complications can be perpetuated, when coupled with economic strain, as lack of financial resources may and often do prevent access to effective medical care (Rolland, 1994a), as was the case with the patient in the Putnam family. According to the VSA Model, the vulnerabilities and stresses associated with SCD interact with one another, and are potentially influenced by the degree to which the patient and family can adapt to and cope with the stresses and vulnerabilities. While the stresses and vulnerabilities of these

families may not differ too much from families with similar types of chronic illness, the ways in which they adapt may greatly differ (Rolland, 1994a, 1994b).

Multiple ways of adapting were evident in this study. The genetic makeup of SCD means that families may need to adapt to, not one, but multiple children with SCD. If fortunate, adaptation may be as fruitful as leading to a "cure" for those families with access to resources – such as financial and educational. The unfortunate delay in proper diagnosis could mean adapting by enduring pain and "faking" wellness for a significant portion of one's life. Participating families converged in the way they joined forces as a family to support one another. They were determined to seek answers and treatment for the patient, as evidenced in this study. The ability of the families to seek support from outside the immediate family system was clearly a strength that contributed to their overall well-being.

Minimizing the losses associated with having SCD was another form of adaptation. Caregivers were mindful to not push the physical limits of the patient, while simultaneously helping the patient lead as much of a "normal" life as possible. Sadly, efforts to provide as normal a life as possible can be easily turned around by the unpredictability of symptom flare-ups. The randomness of symptom flare-ups and the relapsing nature of the illness (i.e., stresses; Karney & Bradbury, 1995) can disturb the daily routine for the patient and their caregivers, whose personal and/or professional lives are often interrupted (Anie, 2005) or placed on hold while they vigilantly wait for another pain crisis (Helps et al., 2003; Wagner et al., 2004). Essentially, families found ways to accommodate to the unpredictability of SCD, thus helping to reduce their stress when symptoms flared up or emergencies happened. This study showed that having contingency plans, which involved immediate or extended family support, as well as

being flexible with their routines and schedules helped participants foster family stability (Karney & Bradbury, 1995).

However, some stresses (e.g., lack of financial resources for the Putnam family) can force maladaptation. This situation may be peculiar to adult patients who have more expectations placed on them by virtue of their maturity compared to children. The lack of understanding and empathy by peers, previous coworkers, and even her extended family, meant that the patient in the Putnam family had to continue on with her daily routines while tolerating excruciating pain. Conversely, pediatric patients can expect to receive more empathy and assistance. Although family systems – immediate and extended – may be willing to help, external systems are not always accommodating or sympathetic, especially to SCD-affected adults (see Strickland et al., 2001). The lack of financial resources and their age can contribute to the degree patients receive emotional support. Given the pressures that adult SCD patients may face, they may be more vulnerable to depression and related psychological conditions when compared to pediatric patients (see Strickland et al., 2001; Thompson et al., 1996).

While difficult and very challenging, the stresses associated with SCD have the potential to draw families – immediate and extended – closer together and to strengthen family ties (Karney & Bradbury, 1995; Rolland, 1994b), just as they can tear them apart (Rolland, 1994b). The findings from this study reflect the former, whereby even in the Putnam family, the patient experienced a better father-daughter relationship (even if it was only in financial and not emotional support) as a result of co-sharing the financial burden of managing SCD. The collective strength of families can reduce the stresses – physical and emotional – of patients and their caregivers. This was the case with the families in this study. The ability of families to

remain stable and functional is highly influenced by their ability to adapt to the combination of family vulnerabilities and life stresses, including managing SCD.

Contributions to Empirical Literature

This study contributes to the body of psychosocial literature on SCD as it is the first to incorporate the voice of SCD patients along with other family members. The collective family unit was invited to take part in this study, so as to give "thick, rich descriptions" (Patton, 2002) of how families have been affected by SCD of a family member. Previous qualitative studies have only elicited the voices of separate members of the family unit (see Hill, 1994a; Mitchell et al., 2007), or the voices of adults patients and their family members separately (see Strickland et al., 2001).

The three families in this study represented a heterogeneous sample. As such, this study sheds more light on the varying family life cycles and how these relate to the extent in which SCD has affected the family unit. At the time of the interview, each family was in a different position than the others. For example, the patient in the Jones family was cured of SCD about 13 years ago. He was a 20-year-old adult at the time of the interview. As he was cured of SCD so long ago, this family is no longer actively adapting to the demands of the illness or having to cope with SCD at all. This family's responses to interview questions were retrospective, as they were no longer actively affected by SCD. Second, the patient in the Whitman family is a child with SCD, who is still affected by this illness, as are his younger sister and mother. While still actively affected by this illness, this family has had time to adapt to the demands of the illness, as the diagnosis was given shortly after the eight-year-old patient was born. Lastly, the patient in the Putnam family and her husband were only informed of the SCD diagnosis about one year ago. While the patient has been combating SCD symptoms and stresses her whole life without

knowing it, she and her husband – and their immediate family unit – are still transitioning to an acceptance of this diagnosis and its prognosis. In addition, they are still actively engaged in the vigilance and the coping strategies that this illness demands.

In other words, each participating family's experiences diverge from one another given each family's unique developmental stage (as outlined above) at the time of the interview. However, regardless of whether the family is currently dealing with SCD or once did many years ago, each family had converging and similar experiences, as are outlined in the three emergent themes above.

Another important contribution this study makes relates to the patient in the Putnam family. This patient was adopted as an infant and therefore did not know her family of origin or her ancestry. Many often mistake her as being "White," though she is actually of Mediterranean descent. Her participation in this study was "to bring awareness" about how pervasive this disease is and that it also affects those of non-African lineages. Her doctors' statement of "But you're White!" is indicative of the perception that exists, even in the medical community, that this illness only affects those of African descent. This misnomer is reflected in the fact that all psychosocial SCD empirical studies found by this researcher have been done on samples or populations identified as being African American or of African ancestry. Future studies on SCD should seek to include those of other lineages as well, as the CDC (2011) has stated that it affects those of African, *as well as* Mediterranean, Indian, Middle Eastern, and Hispanic/Latino descent.

Strengths, Limitations and Further Research

The strength of this study is in the diversity of the families interviewed. Diversity enhances the understanding of the phenomenon by allowing the ability to identify convergence of experiences, as well as to identify reasons for divergence in adaptation.

Limitations of this study include the small sample size. Further studies replicating this study, which included larger family units and extended family members, would help facilitate a broader and more collective experience of the effects of SCD.

The self-selection process of families lends limitation to the study. Those who chose not to participate in this study may not have had similar experiences or were not as cohesive as those who did. If they were not as close or “tight-knit”, they may have been reluctant to share their difficulties with SCD. SCD may have pulled these families apart rather than bringing them together as was the case for the families in this study. Longitudinal studies that begin at the onset SCD, or when SCD is first diagnosed, may provide a better understanding of the factors that contribute to family cohesion and disintegration.

The self-selection process also attracted highly educated families. In each of the participating families, at least one parent or family member had a graduate degree. Participating families are probably not representative of the typical family affected by SCD. Many families may not have access to medical care and resources to know of the latest treatments for SCD (Hill, 1994a). Families may be reluctant to risk the safety of their child and participate in clinical research studies, given their past mistreatment by the government – such as occurred in the Tuskegee Syphilis Study (Tuskegee University, 2008) – and by medical providers, who did not know much about how to care for those with SCD (Hill, 1994b).

The difficulty in recruiting families for this study is reflected in the small sample and the length of time it took to recruit participants. Families who may be the most in need of assistance should to be the focus of future SCD research. Only then can we better understand how families with limited resources, who may be more vulnerable due to their circumstances, adapt to the challenges associated with SCD (see Hill, 1994b).

Another limitation is the lack of responses from participants at the “member checking” (Oktaý, Jacobson, & Fisher, 2013; Patton, 2002) stage. None of the participating families provided feedback to the emergent themes. It is possible that families were in agreement with the final themes. However, without this feedback, it is possible that emergent themes did not, in fact, accurately capture the experiences of all participants. Incentives that encourage “member checking” (Oktaý, Jacobson, & Fisher, 2013; Patton, 2002) could be incorporated in future studies on SCD. This step could help ensure that the voices of participants from minority groups are accurately represented, thus empowering these families (see Charache, Lubin, & Reid, 1989).

With the strengths and limitations of this study having been presented, it is important to note some research questions for future studies. The following research questions are informed by some of the aforementioned limitations of this study and by future research suggestions. The first research question for further study is: *How is family stability affected over time due to the chronic nature of SCD?* This is an important research question to explore as the families in this study were not interviewed or observed longitudinally. The two families who were still affected by SCD of a family member (the Whitman and Putnam families) were interviewed on two occasions which occurred close together. At the time of interviews they indicated that they were tight-knit and that, ultimately, SCD has brought them closer together and made them stronger. However, due to the prognosis of SCD, its accumulative effects and relapsing nature, families may adapt differently over time. Thus, it would be important to explore family relational quality and, subsequently, family stability at much later dates (i.e., one year later, two years later, even five years later) following the initial and follow-up interviews.

Another research question for future study is: *How do families cope and adapt to the demands of SCD if they do not have the support of extended family or community?* To some

degree, each family in this study had the support of immediate and extended family. This support was emotional, physical and/or financial in nature. However, as was illustrated in this study, not all SCD-affected patients are of African lineage. While SCD affects those from predominantly collectivistic cultures (Hurtig, Koepke, & Park, 1989), it cannot be assumed that all SCD patients have immediate or extended family support. In such cases, patients may need to reach out to religious- or spiritually-based support systems, as well as to fictive kin (those who are considered family but are not related by blood). The importance of religion or spirituality as they relate to coping with SCD were never mentioned explicitly by participants in this study. However, these are important resources that are often mentioned as important in the SCD literature (see Strickland et al., 2001; Boykin, Jagers, Ellison, & Albury, 1997).

Clinical Implications

It is important for mental health clinicians (therapists) to be mindful that families' experiences with SCD can be different based upon their social locations and history. Clinicians need to elicit each family's story – how they discovered that their family member has or had SCD, how they are or were affected by SCD, and the progression of the illness with its accompanying stresses. Therapists should not make assumptions about how families are affected by SCD of a family member, though patients and families across cases may be battling similar symptoms.

An important clinical consideration relates to the inherent strengths that many families from collectivist cultures possess – the support from extended families and communities (Boykin et al., 1997; Hill, 1994a). This sense of family and community support can buffer the stresses associated with SCD. Clinicians can link SCD-affected families with SCD support groups or

those for similar chronic illnesses. Such support groups can be an important resource for families (Strickland et al., 2001).

Clinicians can help advocate for patients who have difficulty advocating for themselves and/or communicating with medical staff (McDaniel, Hepworth, & Doherty, 1992). Clinical treatment involving significant family members can be used to identify members who could help advocate for the patient and accompany the patient to medical appointments. It is important for clinicians to be aware of indigent medical care programs and/or case managers in the community. These resources may aid clients in connecting to medical care that they may not otherwise seek out, thus helping to increase their chances of improved physical health and, therefore, psychological health (McDaniel, Hepworth, & Doherty, 1992).

Stressors that affect children also present stressors for the parental/caregiver unit. Conversely, stresses affecting parents/caregivers also affect children (Brown et al., 1993; Campbell, 2003; McDaniel, Hepworth, & Doherty, 1992; Rolland, 1994a, 1994b). Clinicians should offer treatment for the parental/caregiver unit to provide an avenue to process the meaning of having an ill child with a potentially shorter lifespan. The potential loss, ambiguity of what could be lost, and shortened lifespan of a child can be a major stressors for the parental unit (Boss & Couden, 2002).

Consequently, in addition to parental/caregiver units, therapeutic treatment options should include the entire family system. As the majority of populations affected by SCD are collectivistic in nature, interventions focused on the entire family system are likely to be more effective (Hurtig, Koepke, & Park, 1989). Treatment involving the entire family system, that may include extended family and fictive kin, can help prevent the isolation of non-SCD siblings and provide a safe environment to process emotions that may otherwise be overlooked.

While not all families who are affected by SCD of a family member can be compared to one another, therapeutic approaches should strive to find the strengths that each family possesses. For example, a solution-focused treatment approach will seek to find the exceptions and solutions (de Shazer, 1985) to problems that SCD-affected families may already be exhibiting, but may be unable to recognize. In such an approach, less cohesive or struggling families may have difficulty finding exceptions and may share more problem-saturated histories and stories. When this occurs, a narrative approach that focused on meaning-making and challenging problem-focused narratives could prove to be helpful (White & Epston, 1990).

References

- Anie, K. A. (2005). Psychological complications in sickle cell disease. *British Journal of Haematology, 129*, 723-729.
- Barlow, J. H., & Ellard, D. R. (2006). The psychosocial well-being of children with chronic disease, their parents and siblings: An overview of the research evidence base. *Child: Care, Health and Development, 32*, 19-31.
- Barrett, D.H., Wisotzek, I.E., Abel, G.G., Rouleau, J. L., Platt, A. F., Pollard, W. E., et al. (1988). Assessment of psychological functioning of patients with sickle cell disease. *South Medical Journal, 81*, 745-750.
- Belgrave, F. Z., & Molock, S. D. (1991). The role of depression in hospital admissions and emergency treatment of patients with sickle cell disease. *Journal of the National Medical Association, 83*(9), 777-781.
- Boss, P., & Couden, B. A. (2002). Ambiguous loss from chronic physical illness: Clinical interventions with individuals, couples, and families. *Journal of Clinical Psychology, 58*, 1351-1360.
- Boyatzis, R.E. (1998) *Transforming Qualitative Information: Thematic Analysis and Code Development*. Thousand Oaks, London: Sage Publications.
- Boykin, A. W., Jagers, R. J., Ellison, C. M., & Albury, A. (1997). Communalism: Conceptualization and measurement of an Afrocultural social orientation. *Journal of Black Studies, 27*(3), 409-418.
- Brown, R. T., Davies, P. C., Lambert, R., Hsu, L., Hopkins, K., & Eckman, J. (2000). Neurocognitive functioning and magnetic resonance imaging in children with sickle cell disease. *Journal of Pediatric Psychology, 25*, 503-513.

- Brown, R. T., Kaslow, N. J., Doepke, K., Buchanan, I., Eckman, J., Baldwin, K. et al. (1993). Psychosocial and family functioning in children with sickle cell syndrome and their mothers. *Journal of the American Academy of Child and Adolescent Psychiatry*, 32, 545-553.
- Burnes, D. P. R., Antle, B. J., Williams, C. C., & Cook, L. (2008). Mothers raising children with sickle cell disease at the intersection of race, gender, and illness stigma. *Health & Social Work, 33*(3), 211-220.
- Campbell, T. L. (2003). The effectiveness of family interventions for physical disorders. *The Journal of Marital and Family Therapy, 29*, 263-281.
- Centers for Disease Control and Prevention. (2011). Sickle cell disease: Data and statistics. Retrieved from <http://www.cdc.gov/ncbddd/sicklecell/data.html> on March 8, 2014.
- Centers for Disease Control and Prevention. (2008). Sickle cell disease: 10 things you need to know. Retrieved from <http://www.cdc.gov/features/sicklecell/> on December 7, 2008.
- Charache, S. Lubin, B., & Reid, C. D. (1985). *Management and therapy of sickle cell disease*. (NIH Publication No. 85-2117). Washington, D. C.: National Institutes of Health.
- Creswell, J.W. (1998). *Qualitative inquiry and research design: Choosing among five traditions*. Thousand Oaks: Sage Publications.
- Dahl, C. M., & Boss, P. (2005). The use of phenomenology for family therapy research: The search for meaning. In D. H. Sprenkle & F. P. Piercy (Eds.), *Research methods in family therapy* (2nd ed.) (pp. 63-84). New York: Guilford Press.
- Damlouji, N.F., Keves-Cohen, R., Charache, S., Georgopoulos, A., & Folstein, M. F. (1982). Social disability and psychiatric morbidity in sickle cell anemia and diabetes patients. *Psychosomatics, 23*, 925-931.
- de Shazer, S. (1985). *Keys to solution in brief therapy*. New York: Norton.

- Earley, C. J., Kittner, S. J., Feeser, B. R., Gardner, J., Epstein, A., Wozniak, M.A., et al. (1998). Stroke in children and sickle-cell disease: Baltimore-Washington Cooperative Young Stroke Study. *Neurology*, *51*, 169-176.
- Edwards, R., Telfair, J., Cecil, H., Lenoci, J. (2000). Self-efficacy as a predictor of adult adjustment to sickle cell disease: One-year outcomes. *Psychosomatic Medicine*, *63*, 850-858.
- Evans, R. C., Burlew, A. K., & Oler, C. H. (1988). Children with sickle-cell anemia: Parental relations, parent-child relations, and child behavior.
- Famuyiwa, O. O., & Akinyanju, O. O. (1998). Burden of sickle cell anaemia on families of patients. *International Journal of Social Psychiatry*, *44*, 170-180.
- Gil, K., Abrams, G., Phillips, G., & Keefe, F. (1989). Sickle cell disease pain: Relation of coping strategies to adjustment. *Journal of Consulting and Clinical Psychology*, *57*, 725-731.
- Giorgi, A. (1985). *Phenomenology and psychological research*. Duquesne University Press.
- Gold, J. I., Treadwell, M., Weismann, L., & Vachinsky, E. (2008). An expanded transactional stress and coping model for siblings of children with sickle cell disease: Family functioning and sibling coping, self-efficacy and perceived social support.
- Hason, S. P., Hashmi, S., Alhassen, M., Lawson, W., & Castro, O. (2003). Depression in sickle cell disease. *Journal of the National Medical Association*, *95*(7), 533-537.
- Helps, S., Fuggle, P., Udwin, O., & Dick, M. (2003). Psychosocial and neurocognitive aspects of sickle cell disease. *Child and Adolescent Mental Health*, *8*, 11-17.
- Hill, S. A. (1994a). *Managing sickle cell disease in low income families*. Philadelphia: Temple University Press.
- Hill, S. A. (1994b). Motherhood and the obfuscation of medical knowledge: The case of sickle cell disease. *Gender & Society*, *8*, 29-47.

- Hurtig, A. L., Koepke, D., & Park, K. B. (1989). Relationship between severity of chronic illness and adjustment in children and adolescents with sickle cell disease. *Journal of Pediatric Psychology, 14*, 117-132.
- Jenerette, C., Funk, M., & Murdaugh, C. (2005). Sickle cell disease: A stigmatizing condition that may lead to depression. *Issues in Mental Health Nursing, 26*, 1081-1101.
- Karney, B., & Bradbury, T. (1995). The longitudinal course of marital quality and stability: A review of theory, method, and research. *Psychological Bulletin, 118*, 3-34.
- Kim, B. S., Illes, J., Kaplan, R. T., Reiss, A., & Atlas, S. W. (2002). Incidental findings on pediatric MR images of the brain. *American Journal of Neuroradiology, 23*, 1674-1677.
- Lenoci, J., Telfair, J., Cecil, H., & Edwards, R. R. (2002). Development and testing of a tool to access self-care agency in adults with sickle cell disease. *The Western Journal of Nursing Research, 24*(3), 228-245.
- Levenson, J. L., McClish, D. K., Dahman, B. A., Bovbjerg, V. E., Citero, V. D. A., Penberthy, L. T., et al. (2008). Depression and anxiety in adults with sickle cell disease: The PiSCES project. *The Journal of Psychosomatic Medicine, 70*, 192-196.
- McCrae, J. D., & Lumley, M. A. (1997). Health status in sickle cell disease: Examining the roles of pain coping strategies, somatic awareness, and negative affectivity. *Journal of Behavioral Medicine, 21*(1), 35-55.
- McDaniel, S. H., Hepworth, J., & Doherty, W. J. (1992). *Medical family therapy: A biopsychosocial approach to families with health problems*. New York: Basic Books.
- Merriam, S. B. (1998). *Qualitative research and case study applications in education*. San Francisco: Jossey-Bass Publishers.
- Mitchell, M. J., Lemanek, K., Palermo, T. M., Crosby, L. E., Nichols, A., & Powers, S. W. (2007). Parent perspectives on pain management, coping, and family functioning in pediatric sickle cell disease. *Clinical Pediatrics, 46*(4), 311-319.

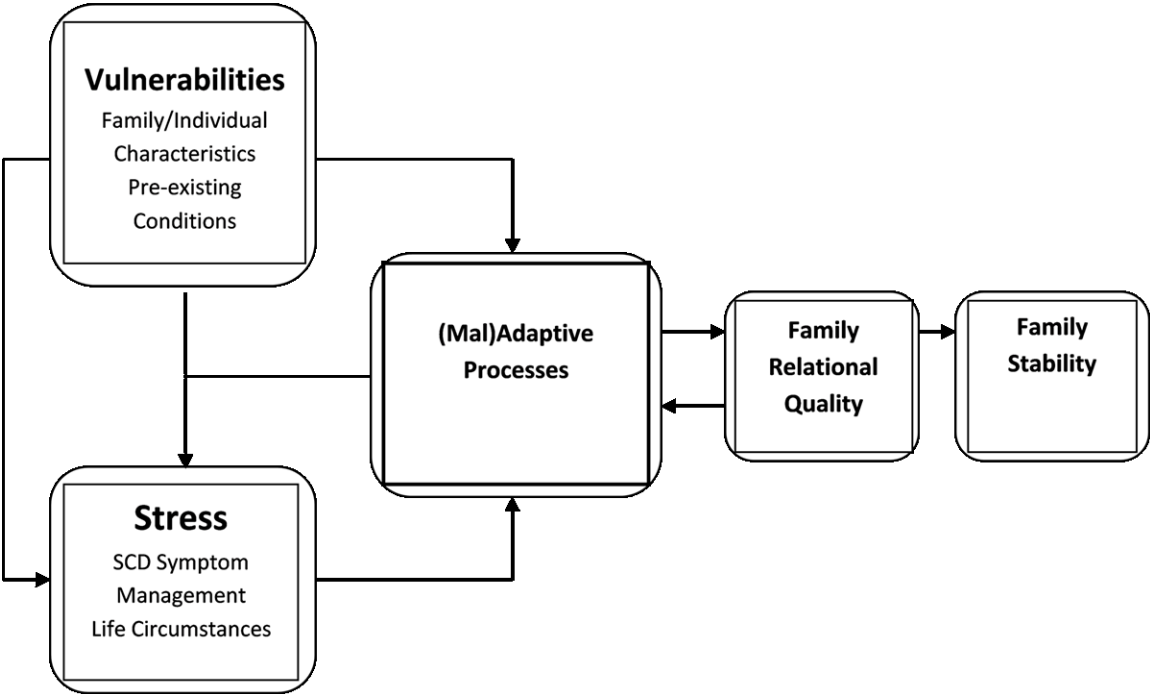
- Morgan, S. A., & Jackson, J. (1986). Psychological and social concomitants of sickle cell anemia in adolescents. *Journal of Pediatric Psychology, 11*, 429-439.
- Nadel, C., Protadin, G. (1977). Psychological factors associated with onset. *New York State Journal of Medicine, 77*, 1075-1078.
- Nishiura, E., & Whitten, C. (1980). Psychosocial problems in families of children with sickle cell anemia. *Urban Health, 9*, 32-35.
- Noll, R. B., Vannatta, K., Koontz, K., Kalinyak, K., Bukowski, W. M., & Davies, W. H. (1996). Peer relationships and emotional well-being of youngsters with sickle cell disease. *Child Development, 67*, 423-436.
- Oktay, J. S., Jacobson, J. M., & Fisher, E. (2013). Learning through experience: The transition from doctoral student to social work educator. *Journal of Social Work Education, 49*(2), 207-221.
- Patton, M. Q. (2002). *Qualitative Research and Evaluative Methods* (3rd ed.). Thousand Oaks: Sage.
- Platt, O. S., Brambilla, D. J., Rosse, W. F., Milner, P. F., Castro, O., Steinberg, M. H., et al., (1994). Mortality in sickle cell disease: Life expectancy and risk factors for early death. *New England Journal of Medicine, 330*, 1639-1644.
- Rolland, J. S. (1994a). *Families, illness, & disability*. New York: HarperCollins.
- Rolland, J. S. (1994b). In sickness and in health: The impact of illness on couples' relationships. *Journal of Marital and Family Therapy, 20*(4), 327-347.
- Schatz, J., McClellan, C. B., Puffer, E. S., Johnson, K., & Roberts, C. W. (2007). Neurodevelopmental screening in toddlers and early preschoolers with sickle cell disease. *Journal of Child Neurology, 23*, 44-50.

- Shapiro, B. S., Dinges, D. F., Orne, E. C., Bauer, N., Reilly, L. B., Whitehouse, W. G., et al. (1995). Home-management of sickle cell-related pain in children and adolescents: Natural history and impact on school attendance. *Pain, 61*, 139-144.
- Sharpe, D., & Rossiter, L. (2002). Siblings of children with a chronic illness: A meta-analysis. *Journal of Pediatric Psychology, 27*(8), 699-710.
- Sickle Cell Disease Association of American, Southern Connecticut, Inc. (2010). About sickle cell disease. Retrieved from <http://www.scdaaofsouthernct.org/> on March 9, 2014.
- Strickland, O. L., Jackson, G., Gilead, M., McGuire, D. B., & Quarles, S. (2001). Use of focus groups for pain and quality of life assessment in adults with sickle cell disease. *Journal of National Black Nurses Association, 12*(2), 36-43.
- Thompson, R. J., Armstrong, F. D., Kronenberger, W. G., Scott, D., McCabe, M. A., Smith, B., Radcliffe, J., Colangelo, L., Gallagher, D., Islam, S., & Wright, E. (1999). Family functioning, neurocognitive functioning, and behavior problems in children with sickle cell disease. *Journal of Pediatric Psychology, 24*, 491-498.
- Thompson, R. J., Gil, K. M., Abrams, M. R., & Phillips, G. (1996). Psychological adjustment of adults with sickle cell anemia: Stability over 20 months, correlates, and predictors. *Journal of Clinical Psychology, 52*(3), 253-261.
- Tuskegee University. (2008). Research ethics: The Tuskegee Syphilis Study. Retrieved from <http://www.tuskegee.edu/global/story.asp?s=1207598&ClientType=Printable> on December 8, 2009.
- Wagner, J. L., Connelly, M., Brown, R. T., Taylor, L. C., Rittle, C., & Wall-Cloues, B. (2004). Predictors of social anxiety in children and adolescents with sickle cell disease. *Journal of Clinical Psychology in Medical Settings, 11*(4), 243-252.
- White, M., & Epston, D. (1990). *Narrative means to therapeutic ends*. New York: Norton.

Wilson Schaeffer, J. J., Gil, K. M., Burchinal, M., Kramer, K. D., Nash, K. B., Orringer, E., et al. (1999). Depression and functioning in relation to health care use in sickle cell disease. *Journal of Behavioral Medicine, 22*, 115-126.

Womeodu, R. J., Bobo-Mosley, L., Brown, C. E., & Gibson, W. R. (1994). In-patient sickle cell mortality clinical characteristics in an urban University Affiliated Hospital The 19th Annual Meeting. The National Sickle Cell Disease Program. New York City, March 23-26, 220.

Appendix A - Vulnerability-Stress-Adaptation Model



The Vulnerability--Stress--Adaptation (VSA) Model (Karney & Bradbury, 1995) model was originally developed to explain the interplay of factors influencing marital quality and stability. For this study, the VSA Model has been extrapolated to family units, rather than marital dyads. Therefore, marital quality and marital stability have been replaced with family relational quality and family stability.

Appendix B - Interview Guide

1. *How does SCD change families?*

Tell me about your family.

Probes:

- How big is your family? Who are the members?
- What are some things do you do together as a family?
- How would you describe your family level of closeness or togetherness?
- What else would you like me to know about your family?

Tell me about when you discovered that you/your family member had SCD?

Probes:

- How were you/your family member informed of the SCD diagnosis?
- How did you/your family react when this was discovered?
- What else was going on with you/your family at that time (birth of a new child, relocation etc.)?
- What has changed the most over time since knowing/discovering that the diagnosis of SCD?
- How has your family's relationships changed since the diagnosis of SCD?
- What has helped your family remain strong?

2. *How do families cope with SCD?*

How have you and your family coped with SCD?

Probes:

- What were some challenges you faced personally and as a family?
- How often does your family talk about SCD? How is it discussed? Who brings it up?
- What have you learned about your family's ability to cope with SCD?
- How has your family's ability to cope changed over time?

3. *How do families adapt to SCD?*

What changes did your family need to make to accommodate SCD?

Probes:

- What have you learned about your family from this experience?
- How has your family's ability to adapt changed over time?
- What made it challenging to adapt?
- For whom were the changes most difficult?

What other things do you feel it would be helpful to share about what it's been like living with and around SCD?

What other things would you like to share that I haven't asked you about?

Appendix C - Follow-up Interview Guide

1. Are there things that you thought of since our previous interview that you want to share or elaborate upon?

2. Is there anything else that you want to share about your family that you would like me to know about?

Probes:

- What have you learned about your family since the patient was diagnosed with SCD?
- (After families indicated how strong or resilient they are) What has helped your family remain strong?

3. After the patient was diagnosed with SCD, what changed the most over time for your family?

Probes:

- What adaptations did you have to make as a family due to the SCD?

Appendix D - Adult Informed Consent Form

Adult Informed Consent Form

K-State IRB #: _____ UNMC IRB #: _____

Title of this Research Study:

**THE EFFECTS OF PEDIATRIC SICKLE CELL DISEASE ON FAMILY
FUNCTIONING: A PHENOMENOLOGICAL STUDY**

Invitation

Your family is invited to take part in this research study. The information in this form is meant to help you decide whether or not to participate in it. If you have questions, please ask them at any time.

Why are you being asked to participate in this research study?

Your family is being asked to take part in this study because you have a child who has sickle cell disease in your family, the child lives in your home, he or she is between the ages of 6-12, and your family (including the patient) speaks English.

What is the reason for doing this research study?

The purpose of this study is to better understand how families with children who have sickle cell disease live with this disease on a daily basis. This study also looks at how family functioning, as your entire family views it, is affected by the child's chronic illness.

What will be done during this research study?

Participation in this study means:

- 1) Your family will be asked to participate in an interview that will last about 1 – 1 ½ hours. This interview will be done in your home at a time that will work for both your family and the researcher. Everyone who is considered as “family” by your child with sickle cell disease is invited to participate.

Initials: _____

- 2) If you participate in the first family interview, a follow-up family interview will be arranged. This will also take part in your home about 1-2 weeks after the first interview. This second interview will only last about 30 minutes. The purpose of this follow-up interview is for your family to share more thoughts and ideas about how your family sees it is affected by sickle cell disease that were not thought of during the first interview.
- 3) All interviews will be audiotaped and videotaped, then transcribed. This will help the researcher remember what you said during the first and follow-up family interviews. This also will help the researcher with the purpose of the study, to see how your family functioning is affected by pediatric sickle cell disease.

What are the possible risks of being in this research study?

Your family or family members may experience some emotional discomfort when answering questions about the child's sickle cell disease and how it has affected the family. There is a possibility for loss of confidentiality.

What are the possible benefits of this research study for you?

The family interviews will allow your family the opportunity to talk about its experiences with sickle cell disease. However, you may not get any benefit from being in this study.

What are the possible benefits to other people?

By participating in this study, you will help other people who are affected by sickle cell disease know that they are not alone and have had similar experiences. Those who do not have a child with sickle cell disease will better understand what it is like to have a young family member with this disease. The results of this study will eventually help to create a counseling treatment approach that will help families affected by childhood sickle cell disease, and other chronic illnesses.

What are the alternatives to being in this research study?

Instead of being in this study, your family or individual family members can choose not to participate.

What will being in this research study cost you?

There is no cost to you for being in this research study.

Initials: _____

Will you be paid for being in this research study?

Your family will be paid \$50 in the form of a gift card for its participation in this study. \$50 (in the form of a gift card) will be paid to those families who complete at least the first of two interviews.

What should you do if you have a problem during this research study?

Your welfare is the major concern of the researcher of this study. If you have a problem as a direct result of being in this study, you should immediately contact and speak to the researcher. His contact information is located at the end of this form. You will be given a copy of this form for your records.

How will information about you be protected?

Reasonable steps will be taken to protect your privacy and the confidentiality of your study data. An announcement will be made at the beginning of the interviews that your interviews are strictly confidential. Your names will be kept confidential and instead of your names codes will be used. Transcripts will only use codes and not your real names. A code key will be used only by the researcher in helping him to transcribe interviews. Transcripts will not use any identifying information. All audio- and videorecordings will be kept locked in the researcher's file cabinet when not in use for up to 5 years. After that, all recordings will be destroyed. The only person who will have access to your research data is the researcher, the Institutional Review Board (IRB), and any other person or agency required by law. The information from this study may be published in scientific journals or presented at scientific meetings, but your identities will be kept strictly confidential. The researcher plans to share his research findings with you to ensure that they accurately reflect your true, lived experiences with sickle cell disease.

What are your rights as a research subject?

You have rights as a research subject. Your rights have been explained to you in this consent form and in The Rights of Research Subjects that you have been given. If you have any questions concerning your rights or complaints about the research, talk to the researcher or contact the IRB by:

- Telephone (402) 559-6463
- E-mail: IRBORA@unmc.edu
- Mail: UNMC Institutional Review Board, 987830 Nebraska Medical Center, Omaha, NE 68198-7830

Initials: _____

What will happen if you decide not to be in this research study or decide to stop participating once you start?

Your family or family members can decide not to be in this research study or your family or family members can stop being in this research study (“withdraw”) at any time before, during, or after the research begins. Deciding not to be in this study or deciding to withdraw will not affect your relationship with the researcher, The University of Nebraska Medical Center (UNMC), University Medical Associates (UMA), or your physician.

Your family or family members will not lose any medical benefits to which you are entitled. If the research team gets any new information during this research study that may affect whether you would want to continue being in the study, you will be informed promptly.

Documentation of informed consent

Each family member, please indicate your decision below and initial and date each one.

Each family member please check **YES** if you agree to participate in the first and follow-up interviews about your experiences living with and around childhood sickle cell disease on a daily basis. Each family member can check **NO** if you do not agree to participate in one or both interviews. If needed, additional family members can write YES or NO on the back of this page and initial and date next to their response.

#1 YES _____	NO _____	_____	_____
		Initials	Date
#2 YES _____	NO _____	_____	_____
		Initials	Date
#3 YES _____	NO _____	_____	_____
		Initials	Date
#4 YES _____	NO _____	_____	_____
		Initials	Date
#5 YES _____	NO _____	_____	_____
		Initials	Date
#6 YES _____	NO _____	_____	_____
		Initials	Date
#7 YES _____	NO _____	_____	_____
		Initials	Date

Initials : _____

You are freely making a decision whether to be in this research study. Signing this form means that you (1) have read and understand this consent form, (2) have had the consent form explained to you, (3) have had your questions answered, and (4) have decided to be in the research study. If necessary, additional family members can print their names, sign, date, and list the time they do so on the back of this page.

If you have any questions during the study, you should talk to the researcher listed below. You will be given a copy of this consent form to keep.

#1 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#2 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#3 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#4 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#5 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#6 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#7 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#8 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

#9 Printed name of Subject: _____

Signature of Subject: _____ Date: _____ Time: _____

Initials: _____

My signature below certifies that all the elements of informed consent described on this consent form have been explained fully to the subjects. In my judgment, the participants possess the legal capacity to give informed consent to participate in this research and are voluntarily and knowingly giving informed consent to participate.

Signature of Person Obtaining Consent

Date

Authorized Study Personnel

Principal Investigator/Researcher

Kevin C. Garrett, M.Ed., Doctoral Student and MFT Intern
Kansas State University and Department of Family Medicine, UNMC
Office: (402) 559-8072

Secondary Investigators

W. David Robinson, PhD, LIMHP
Department of Family Medicine, UNMC
Office: (402) 559-5868

Tony P. Jurich, PhD, LCMFT
Kansas State University
Office: (785) 532-1488

Joyce Baptist, PhD, LCMFT
Kansas State University
Office: (785) 532-6891

Initials: _____

Appendix E - Parental Consent Form



COLLEGE OF MEDICINE
Department of Family Medicine
Research Division

UNMC IRB # _____

K-State IRB # _____

PARENTAL CONSENT FORM

Title of This Research Study: THE EFFECTS OF PEDIATRIC SICKLE CELL DISEASE ON FAMILY FUNCTIONING: A PHENOMENOLOGICAL STUDY.

Invitation

Your child/children is/are invited to participate in a research study. The following information is provided in order to help you to make an informed decision whether or not to allow them to participate. If you have any questions, please call Kevin C. Garrett, M.Ed., PLMHP, who is the principal investigator, at 402-559-8072.

Why is your child being asked to be in this research study?

Your child is eligible to participate in this study because he or she has sickle cell disease, is between the ages of 6-12, and speaks English. Other children and siblings are invited to attend because they with the child with sickle cell disease.

What is the reason for doing this research study?

The purpose of the study is to better understand how families with children who have sickle cell disease live with this disease on a daily basis. This study also looks at how family functioning, as your entire family (including your child with sickle cell disease) views it, is influenced by the child's illness. As part of the entire family, other children in your home who are 6 years old and older also are invited to participate in the study.

Initials: _____

What will be done during this research study?

Participation in this study will involve completing 2 interviews: (1) a first interview which will last about 2 hours and (2) a follow-up interview which will last about 30 minutes. Some questions will ask about the age of your child when he or she was diagnosed with sickle cell disease and other questions will ask about your family. The 2 interviews will be done in your home or a convenient, private site for your family. Both interviews will be videotaped and audiotaped, and then these interviews will be typed (“transcribed”).

What are the possible risks of being in this research study?

The potential risks to study subjects are minimal. Your child or family may experience some emotional discomfort when answering questions about the child’s sickle cell disease. There is the possibility for loss of confidentiality because interviews are being done in your home or another private location.

What are the possible benefits to your child?

There is the potential that your child will not benefit from his or her participation in the study.

The possible benefit for your child may include the opportunity for him or her to share his or her experiences living with sickle cell disease.

What are the possible benefits to other people?

Your child’s participation in this study has the potential to help other people affected by sickle cell disease to know that they are not alone and that others have had similar experiences. The results of this study will eventually help to create a counseling treatment approach that will help families affected by childhood sickle cell disease and other chronic illnesses.

What are the alternatives to being in this research study?

Instead of being in this research study, your child can choose not to participate. Deciding not to participate in the study will not affect your child’s relationship with the researcher, The University of Nebraska Medical Center, University of Nebraska Medical Center Physicians, or his or her doctor.

Initials: _____

What will being in this research study cost your child?

There is no cost to your child or family to be in this research study.

Will your child be paid for being in this research study?

Your child and family will be given a \$50.00 gift card after you complete at least the initial interview. Your child and family will need to at least complete the first of the two interviews to be paid the gift card.

What should your child do if he or she has a problem during this research study?

Your child’s welfare is the major concern of every member of the research team. If your child has a problem as a direct result of being in this study, he or she should immediately contact one of the people listed at the end of this consent form.

How will information about your child be protected?

Reasonable steps will be taken to protect your child’s and family’s privacy and the confidentiality of study data. An announcement will be made at the beginning of the interviews that your interviews are strictly confidential. Participants will be assigned a code at the beginning of the study. All data collected from each participant will be identified only by code. Interview transcripts will not use any identifying information; they will solely contain the codes. All audio- and videorecordings will be kept in the researcher’s locked file cabinet.

Contact information of participants will be used to obtain informed consents and contact the participants for follow-up data collection. This personal contact information will be stored separately from the actual data.

The only persons who will have access to your child’s research records are the study personnel, the Institutional Review Board (IRB), and any other person or agency required by law. The information from this study may be published in scientific journals or presented at scientific meetings but your child’s identity will be kept strictly confidential.

Limits to Confidentiality: As mentioned above, steps will be taken by the researcher to try to keep your child’s and family’s information confidential. However, there are several reasons why the researcher may have to break confidentiality. These reasons include: (1) suspected or reported child abuse/neglect, elder abuse/neglect, and/or vulnerable population abuse/neglect; (2) your child or a family member says that he or she is going to harm someone else or harm themselves; and (3) when ordered by a court of law.

Initials: _____

What are your child’s rights as a research subject?

Your child has rights as a research subject. These rights have been explained in this assent form and in *The Rights of Research Subjects* that your child has been given. If you have any questions concerning your rights, talk to the investigator or call the Institutional Review Board (IRB), telephone (402) 559-6463.

What will happen if your child decides not to be in this research study or decides to stop participating once he/she starts?

Your child can decide not to be in this research study, or your child can stop being in this research study (“withdraw”) at any time before, during, or after the research begins. Deciding not to be in this research study or deciding to withdraw will not affect your child’s relationship with the researcher, The University of Nebraska Medical Center, University of Nebraska Medical Center Physicians, or your doctor.

Your child will not lose any benefits to which he/she is entitled.

If the research team gets any new information during this research study that may affect whether your child would want to continue being in the study you and your child will be informed promptly.

Documentation of informed assent

You are freely making a decision whether to allow your child to be in this research study. Signing this form means that (1) you have read and understood this consent form, (2) you have had the consent form explained to you, (3) you have had your questions answered and (4) you have decided to allow your child to be in the research study.

If you have any questions during the study, you should talk to one of the investigators listed below. You will be given a copy of this consent form to keep.

Initials _____

SIGNATURE OF PARENT

DATE

TIME

PRINTED NAME OF CHILD

SIGNATURE OF INVESTIGATOR

DATE

Authorized Study Personnel:

Principal Investigator

Kevin Garrett, M.Ed., PLMHP
(402) 559-8072

Secondary Investigators

W. David Robinson, PhD, LIMHP
(402) 559-5868

Tony P. Jurich, PhD, LCMFT
(785) 532-1488

Joyce Baptist, PhD, LCMFT
(785) 532-6891

Initials: _____

Appendix F - Child Assent Form



COLLEGE OF MEDICINE
Department of Family Medicine
Research Division

UNMC IRB # _____

K-State IRB # _____

CHILD ASSENT FORM

Title of This Research Study: THE EFFECTS OF PEDIATRIC SICKLE CELL DISEASE ON FAMILY FUNCTIONING: A PHENOMENOLOGICAL STUDY.

I am doing a study because I want to learn more about what it is like for you and your family to live with sickle cell disease every day. I am asking for your help because I don't know very much about what it's like for kids and the whole family to live with sickle cell disease every day.

If you agree to be in my study, I am going to ask you some questions about you and your family about themselves. I am going to ask you questions about how your family talks, helps one another, and what you and they think about sickle cell disease.

You can ask me questions at any time. If you decide that you don't want me to ask you questions, you don't have to start the study. If you decide at any time not to finish answering questions, you can ask me to stop. The questions I ask you and your family are about what you and they think. There are no right or wrong answers, because I want to know what you and your family think about things.

If you sign this paper, it means that you have read this and I have talked to you about it and answered any questions. If you sign this, it means that you want to be in the study. If you don't sign it, it means that you don't want to be in the study. No one will be mad if you decide not to be in the study or if you decide to stop being in the study once you start.

SIGNATURE OF CHILD

DATE

TIME

PRINTED NAME OF CHILD

SIGNATURE OF INVESTIGATOR

DATE

PRINTED NAME OF INVESTIGATOR

Authorized Study Personnel:

Principal Investigator

Kevin Garrett, M.Ed., PLMHP
(402) 559-8072

Secondary Investigators

W. David Robinson, PhD, LIMHP
(402) 559-5868

Tony P. Jurich, PhD, LCMFT
(785) 532-1488

Joyce Baptist, PhD, LCMFT
(785) 532-6891

Initials: _____

Appendix G - Rights of Research Subjects Handout



Institutional Review Board (IRB)

THE RIGHTS OF RESEARCH SUBJECTS

AS A RESEARCH SUBJECT AT THE NEBRASKA MEDICAL CENTER

YOU HAVE THE RIGHT ...

... to be told everything you need to know about the research before you are asked to decide whether or not to take part in the research study. The research will be explained to you in a way that assures you understand enough to decide whether or not to take part.

... to freely decide whether or not to take part in the research.

... to decide not to be in the research, or to stop participating in the research at any time. This will not affect your medical care or your relationship with the investigator or the Nebraska Medical Center. Your doctor will still take care of you.

... to ask questions about the research at any time. The investigator will answer your questions honestly and completely.

... to know that your safety and welfare will always come first. The investigator will display the highest possible degree of skill and care throughout this research. Any risks or discomforts will be minimized as much as possible.

... to privacy and confidentiality. The investigator will treat information about you carefully, and will respect your privacy.

... to keep all the legal rights you have now. You are not giving up any of your legal rights by taking part in this research study.

... to be treated with dignity and respect at all times

The Institutional Review Board is responsible for assuring that your rights and welfare are protected. If you have any questions about your rights, contact the Institutional Review Board at (402) 559-6463.

Appendix H - Outline of Emergent Themes and Sub-themes

Overarching Research Question:

How are family relational quality and family stability affected by SCD?

Sub-research Questions:

1. *What challenges does SCD pose to families?*
2. *How do families adapt to and cope with the demands of SCD?*
3. *What contributes to family relational quality and stability?*

Theme #1: *Stress and Challenges*

Sub-themes:

- *SCD Symptoms*
- *Non-SCD symptoms and stresses*

Theme #2: *Adapting to and Coping with the Demands of SCD*

Sub-themes:

- *Education and Acceptance*
- *The Necessity of Medical Care*
- *Family Support and Family Time*
 - *Extended Family*
 - *Family Time*

Theme #3: *Individual and Family Strengths*

Appendix I - Participating Family Demographics Summary

The Whitman Family

- From the Midwest region of the U.S.
- African American
- SCD patient is male, was 8 years old at time of interview
- SCD patient has younger 3-year-old sister who has SCD
- Patient's mother has a master's degree and is a helping professional
- Patient's father incarcerated at time of interview
- SCD patient and his mother first interviewed in home of maternal grandparents
- Patient's mother (without patient) completed phone conference follow-up interview 2 weeks after initial face-to-face interview
- Initial and follow-up interviews took 2 hours combined to complete

The Jones Family

- From the Southern region of the U.S.
- African American
- Patient was 20 years old at time of interview; patient was 7 years old when cured of SCD through a bone marrow transplant
- Patient's father is a doctor
- Patient's mother has a master's degree
- Patient has younger teenage sister who does not have SCD
- Patient with both of his parents first interviewed via Skype
- Patient with both of his parents completed phone conference follow-up interview 3 weeks after initial interview
- Initial and follow-up interviews took 1.5 hours combined to complete

The Putnam Family

- From the Eastern region of the U.S.
- Patient is of Mediterranean descent; appears "White"
- Patient was adopted as an infant; adopted father did not want to know about her family of origin
- Patient is a 35-year-old female
- Patient diagnosed with SCD 1 year ago
- Patient has master's degree and is a helping professional
- Patient has teenage son who declined participation in the study
- Patient and her husband were first interviewed via Skype (no video, only audio)
- Patient and her husband completed phone conference follow-up interview 4 weeks after initial interview
- Initial and follow-up interviews took 2 hours combined to complete