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A Case-Control Study: The Psychosocial Functioning and Academic Achievement in Siblings with and without Sickle Cell Disease

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ABSTRACT

A Case Control Study: The Psychosocial Functioning and Academic Achievement in Siblings with and without Sickle Cell Disease

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Objective: The aim of this study was to compare the psychosocial functioning and academic achievement in siblings with and without sickle cell disease.

Methods: Using convenience sampling, we recruited (N=133) 45 siblings with sickle cell disease, 43 siblings without sickle cell disease, and 45 primary caregivers from the Children's National Medical Center Sickle Cell Program in Washington, DC. We controlled for age, family environment, socioeconomic status, and parent education in the dyad. Siblings with and without sickle cell disease, ages 12-18, completed the Youth Self Report. Their primary caregivers completed the Child Behavior Checklist for each sibling. Demographic information was gathered based on primary caregivers' responses to a general information data sheet.

Results: Siblings with sickle cell disease (M=53.70, SD=9.01) reported more internalizing behaviors than their healthy siblings (M=46.51, SD=7.73), t=4.52, p=0.00. They also reported less social competence (M=43.52, SD=9.55) than their healthy siblings (M=50.38, SD=7.23), t= 3.78, p=0.00. Primary caregivers reported similar results. They reported more internalizing behaviors in children with sickle cell disease (M=54.96, SD=8.45) than in children without sickle cell disease (M=47.31, SD=8.16), t=4.83, p=0.00. However, they reported less social competence in children without sickle

cell disease (M=52.11, SD=3.96) than in children with sickle cell disease (M= 42.20, SD=13.75), t=-4.71, p=0.00. And lastly, primary caregivers reported higher academic achievement in children with sickle cell disease (M=42.16, SD=11.20) than in children without sickle cell disease (M=50.82, SD=7.39), t=-4.55, p=0.00.

Conclusion: This study provides additional evidence that adolescents with sickle cell disease are at risk for psychosocial adjustment problems and poor academic achievement. It also provides evidence that adolescents with sickle cell disease can cope as well as their healthy siblings with adequate family support, as all subjects had involved families and all children with sickle cell disease were involved in treatment. More longitudinal studies are needed to understand the impact of sickle cell disease from adolescence to adulthood. This study highlights the need for additional resources and interventions to address the needs of this particular patient population.

Keywords: psychosocial, adolescents, sickle cell disease, academic achievement

DEDICATION

I dedicate this dissertation to my family for their love, support, and encouragement.

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CHAPTER 1

Introduction

In the United States, the number of children and adolescents with chronic illnesses has increased drastically over the past 30 years (Callahan, Winitzer, & Keenan, 2001). The current data suggest that 7.5 million children and adolescents have a chronic illness (Thompson & Gustafson, 1996). Twenty percent of them have a mild chronic illness, 9% have a moderate chronic medical illness, and 2% have a severe chronic illness (Reiss & Gibson, 2010). Thus more than 1 million children have a severe chronic illness that may impair their daily functioning (Thompson & Gustafson, 1996). Furthermore, it is estimated that 31.5 % or 1 in 10 adolescents in the United States have a chronic illness (Callahan et al., 2001; Thompson & Gustafson, 1996).

The Commission on Chronic Illness defines chronic illness as "all impairments or deviations from normal which have one or more of the following characteristics: are permanent, leave residual disability, are caused by non-reversible pathological alterations, requiring training of the patient for rehabilitation, and may be expected to require a long period of supervision, observation or care" (Royer, 1998, p. 1). Mattson (1972) defines chronic illness as "a disorder with a protracted course which can be progressive and fatal or associated with a relatively normal life span despite impaired physical or mental functioning" (p. 801). The illness usually involves one or more organs and impairs both health and psychological functioning (Sieh, Meijer, Oort, Visser-Meily, & Leij, 2010). Generally, it is expected to last, at the time of diagnosis, at least 12 months and is usually characterized by periods of exacerbation, remissions and degeneration (Dobbie & Meller,

2008; Royer, 1998).

Adolescents diagnosed with a chronic illness are at risk for psychosocial difficulties. Numerous studies have shown that adolescents with chronic illness are at higher risk for psychosocial difficulties compared to healthy adolescents (Hurtig & White, 1985; Moise, 1986; Morgan & Jackson, 1986; Wallander, Varni, Babani, Banis, & Wilcox, 1988; Rodrigue, Streisand, Banko, Kedar, & Pitel, 1996). The most reported psychosocial problems reported in the literature have included poor self-concept, increased anxiety and depression, social withdrawal or aggression, and maladaptive family and peer relationships (Hurtig & White, 1985; Moise, 1986; Morgan & Jackson, 1986; Wallamder, Varni, Babani, Banis, & Wilcox, 1988; Rodrigue et al., 1996). In addition, lower educational achievement, more frequent truancy, and psychiatric disorders have been reported in the literature (Hurtig & White, 1985; Moise, 1986; Morgan & Jackson, 1986; Wallander et al., 1988; Rodrigue et al., 1996)). Symptoms of anxiety, including feeling tense, worried, and fearful, have been estimated at about 7-40% for adolescents with a chronic illness (Pao & Bosk, 2010). These psychological problems have been identified in adolescents with various chronic illnesses, including HIV, diabetes, cancer, juvenile rheumatoid arthritis, and asthma. The severity of these symptoms depends on various variables such as social support system, race, age and socioeconomic status. Although most adolescents with chronic illness function quite well despite the many difficulties and adversities associated with their illness, some adolescents are at risk for psychosocial dysfunction.

Sickle cell disease is a chronic illness that affects more than 250 million people

worldwide (Bediako, Lavender, & Yasin, 2007). In the United States, it affects 80,000 to 100,000 Americans or 0.25% of the population (Consensus Conference, 1987). It is most common among people of African, Caribbean, Central and South American, and Mediterranean decent. One in 12 African Americans is a carrier, and 1 in 400 African Americans have the disease (Charache, Luban, & Reid, 1991; Hurtig, Koepke, & Park, 1989; Lemanek, Buckloh, Woods, & Bulter, 1995). It affects approximately 1 in every 400 to 500 African American newborns (Barakat, Patterson, Weinberger, Simon, Gonzalez, & Dampier, 2007; Hurtig et al., 1987) and 1 in every 19,000 Latino American newborns (Driscoll, 2009). It is one of the most common genetic disorders in the United States and the most common genetic disorder in the world (Edwards et al., 2005; Kelch-Oliver, Smith, Diaz, & Collins, 2007).

Sickle cell disease can have a profound impact on adolescents' psychological development. Self-esteem issues and poor body image due to delayed growth and sexual maturation may cause significant psychological distress (Tanyi, 2003). Frequent pain crises, leg ulcers from poor wound healing and avascular necrosis of the hip and shoulder are significant threats to adolescents' self-concept (Tanyi, 2003). Problems with parental and peer relationships are also sources of psychological distress (Tanyi, 2003). In addition, frequent hospitalizations and clinic appointments may affect an adolescent's ability to become independent of parents and family members. As a result, poor psychological adjustment in adolescents with sickle cell disease may lead to low self-esteem and poor academic performance and achievement.

The aim of this study is to compare the psychosocial functioning and academic

achievement in siblings with and without sickle cell disease. This quantitative study will use a case control design. Using convenience sampling, this researcher will recruit (N=150) 50 siblings with sickle cell disease, 50 siblings without sickle cell disease, and 50 caregivers from the Children's National Medical Center Sickle Cell Program in Washington, DC. This researcher will control for age, family environment, socioeconomic status, and parent education in the dyad. Siblings with and without sickle cell disease will complete the Youth Self Report. Primary caregivers will complete the Child Behavior Checklist for each sibling.

This study will address the following research question: to what extent do siblings with and without sickle cell disease differ in psychosocial functioning and academic achievement? This study hypothesis is that psychosocial functioning and academic achievement will be lower in siblings with sickle cell disease than in siblings without sickle cell disease.

Results from previous research examining the psychosocial functioning and academic achievement of adolescents with sickle cell disease have shown that adolescents with sickle cell disease are at increased risk for psychosocial adjustment problems and poor academic achievement. However, many of these were poorly designed and used healthy populations or other chronic disease populations as benchmark comparisons. In addition, there is considerable variability in etiology, manifestation, and treatment regimen among chronic diseases. Other studies used clinical narratives or observations, with no standard measures of behaviors and no comparison with test norms or controls. Furthermore, many of these studies were conducted more than 20-years ago

and are outdated. And lastly, many of these studies were conducted with low-income children and adolescents (Casey, Brown, & Bakerman, 2000; Hurtig & Parker, 1989). However, the Children's National Medical Center Sickle Cell Program serves children and adolescents from various socioeconomic backgrounds.

As a result, there is compelling evidence to support a well-designed study that compares the psychosocial functioning and academic achievement in siblings with and without sickle cell disease. This researcher seeks to understand the impact of sickle cell disease on psychosocial functioning and academic achievement by comparing siblings with and without sickle cell disease. This study will improve on previous research by using a case control design that has higher internal validity. This study will use convenience sampling to recruit a large sample of siblings with and without sickle cell disease from the Washington, DC metropolitan area. Lastly, this study will control dyads for age, socioeconomic status, family environment, and parent education.

This is an important study to conduct because poor psychosocial adjustment and academic achievement in adolescents with sickle cell disease is associated with poor medical outcomes, high levels of psychopathology, and poor quality of life. As a result, this study is expected to lead to a greater understanding of the psychosocial and academic needs of adolescents with sickle cell disease and lead to the development and implementation of additional resources and interventions to address the needs of this particular patient population.

CHAPTER 2

The Pathophysiology of Sickle Cell Disease

Sickle cell disease is a chronic condition that is characterized by the predominance of hemoglobin S in red blood cells (Gold, Mahrer, Treadwell, Weissman, & Vichinsky, 2008). It is estimated that 70% to 95% of the hemoglobin in patients with sickle cell disease is abnormal (Tany, 2003). Hemoglobin is essential for the transport of oxygen throughout the body. It is composed of two alpha globulin chains and two beta globulin chains (Gold et al, 2008). In sickle hemoglobin, the beta globulin is different from normal globulin by the replacement of valine (Gold et al., 2008). This substitution of valine causes the hemoglobin S molecules to deoxygenate. As a result, the oxygen-carrying capacity of the red blood cell is significantly less in patients with sickle cell disease.

The average life span of a healthy red blood cell is 120 days. However, in patients with sickle cell disease the life span is 10 to 20 days. Their red blood cells are crescent or sickle shape, rigid and sticky and have difficulty passing through tiny vessels in the body (Gold et al., 2008; Koch, Yang, Olney, 2000). As a result, patients are at increased risk for destruction of blood vessels, impaired circulation, increase blood viscosity, decreased perfusion to organs, and tissue hypoxia, infarction, and necrosis (Tanyi, 2003). They may experience acute painful episodes or crises, multi-organ and system failure, and neurological complications (Driscoll, 2009; Edwards et al., 2005).

There are five main sickle cell genotypes. They are sickle cell anemia (Hb SS),

sickle cell-hemoglobin C disease (Hb SC), sickle cell β thalassemia, which includes sickle cell ß+ thalassemia and sickle cell ß° thalassemia (Hb SB O thal), Hb SO arab, and SD Punjad (Driscoll, 2009; Koch, et al., 2000; Platt & Sacerdote, 2002). While patients with sickle cell ß +thalassemia have approximately 3- 25% Hb A, patients with sickle cell ß° thalassemia have an absence of Hb A. Generally, patients with sickle cell ß+ thalassemia have a fairly benign clinical course compared to patients with sickle cell ß° thalassemia (Koch et al., 2000; Tanyi, 2003). Similarly, patients with sickle cell hemoglobin C disease usually have a fairly mild clinical course (Koch et al., 2000; Tanyi, 2003).

Sickle cell anemia (Hb SS) is the most common genotype. It affects approximately 1 in 375 African Americans (McCrae & Lumley, 1998). These people typically have lower hemoglobin levels and experience earlier, more frequent and more severe symptoms than the other genotypes (Bonner, Gustafson, Schumacher & Thompson, 1999). They account for a significant number of the hospitalizations in people with sickle cell disease.

Patients with sickle cell disease are at significant risk for serious pathophysiologic complications. Such complications are often unpredictable and may include chronic anemia, vaso-occlusive crises (severe pain), stroke, infections (especially pneumococcal), acute chest syndrome (a potentially life-threatening condition characterized by chest pain, fever, and pneumonia-like symptoms), pulmonary embolus, pulmonary hypertension, cholecystitis, splenic sequestration (enlarged spleen due to poor drainage and pooling of the blood, which may require surgical removal of the spleen), retinal detachment and

hemorrhage, and aseptic necrosis of the joints (Driscoll, 2009; Lutz, Barakat, Smith-Whitley, & Ohene-Frempong, 2004). Sickle cell disease may tax the cardiovascular system, resulting in reduced exercise tolerance. Delays in growth and sexual development also occur, and males may experience priapism or painful, undesired erection lasting for more than 12 hours (Barakat et al., 2007; Driscoll, 2009). Patients with sickle cell disease are at increased risk for neurological disease or stroke. The most common causes of stroke in patients with sickle cell disease are cerebrovascular infarction and neurological disease (Driscoll, 2009; Gil, Williams, Thompson, & Kinney, 1991; Lemanek et al., 1995; Ware, Zimmer, & Schultz, 1999). These complications are often life-long and tend to increase in severity with age (Kedar & Pitel, 1996; Kramer, Rooks, Washington, & Pearson, 1980)

Vaso-occlusive crisis

One of the most common complications of sickle cell disease is unpredictable and recurrent pain. The pain is often widespread and migratory. It is generally bilateral and symmetrical and it may move from one joint to another joint (Jacob, 2001). Tenderness and pressure are common as well as swelling of the affected area (Jacob, 2001). The pain is often “bone pain” and commonly involves the lower back, chest, femoral shaft and hip joints, ribs, knees, abdomen, and head. Abdominal pain is one form of acute pain. It is often associated with distention and ileus (Jacob, 2001). The pain may result in removal of the gallbladder and appendix. Abdominal, chest, and lower back pain are common. Priapism, which is caused by sickling in the sinusoids of the penis, results in painful erection that may last for hours or days or become chronic. It is one of the most feared

and difficult sickle cell conditions to treat. In severely affected patients, it commonly results in a loss of sexual function. And lastly, some people experience pain crises with such frequency as to become severely disabled.

Medical management of sickle cell pain is primarily aimed, first, at treating the underlying medical condition by minimizing the factors associated with intravascular sickling and, second, at treating the pain as effectively and efficiently as possible (Yaster et al., 2000). This may include the use of opioids such as morphine and hydromorphone, non-steroidal anti-inflammatory agents such ibuprofen or ketorolac, hydration, physical therapy, and ancillary therapies such as relaxation or guided therapy (Yaster et al., 2000). Often, the goal of treatment is to minimize the frequency and duration of hospitalizations and emergency room visits and facilitate home management of pain. The average length of hospitalization for a vaso-occlusive crisis is 4 days. According to the research, vaso-occlusive crisis is an indicator of clinical severity and correlates with early mortality in patients with sickle cell disease (Yaster et al., 2000).

Infection

Another complication of sickle cell disease is bacterial infections. Bacterial infections are a major cause of morbidity and mortality in patients with sickle cell disease. Such organisms include Streptococcus pneumoniae, Hemophilus influenzae, Neisseria meningitidis, Salmonella species, Mycoplasma pneumoniae, Staphyloccus aureua, Escherichia coli, and Streptococcus pyogenes (National Institutes of Health, 1996). Streptococcus pneumoniae is 400 times higher in patients with sickle cell disease than in healthy individuals (Driscoll, 2009). Septicemia/ meningitis is the main cause of

death in early childhood. Increased risk of bacterial infections continues into adolescence and adulthood in patients with hemoglobin C disease, hemoglobin SS disease, and sickle beta thalassemia.

Infections, whether bacterial, viral (rubeola or cytomegalovirus), granulomatous (tuberculosis, coccidioidomycosis, histoplasmosis), or parasitic (malaria) cause greater morbidity, spread more rapidly, and are more difficult to treat in patients with sickle cell anemia (National Institutes of Health, 1996). Certain infections may lead to life-threatening exacerbations of anemia. They may also precipitate vaso-occlusive crises in patients. Human parvovirus is an infection that is common in patients with sickle cell disease (National Institutes of Health, 1996). Prevention or early aggressive treatment of infection is one of the most important aspects of treatment in these patients.

Auto-infarction

A major complication of vaso-occlusion in early childhood is self-destruction or auto-infarction of the spleen. By 1 year of age, approximately 30% of children who have sickle cell anemia have impaired splenic functioning (National Institutes of Health, 1996). However, by 6 years of age, 90% of children who have sickle cell anemia have impaired splenic functioning (National Institutes of Health, 1996). The spleen is an organ that filters germs, thereby protecting people against infection. During the process of auto-infarction, the spleen initially enlarges before shrinking to a dysfunctional mass (National Institutes of Health, 1996). Another cause of splenic enlargement is splenic sequestration. In this situation, the spleen enlarges due to the trapping of red blood cells.

The spleen seldom burst in this situation, but the patient's blood becomes confined to the spleen, causing shock. Thus, splenic sequestration further increases patients' risk of infection.

Acute Chest Syndrome and Fat Embolization Syndrome

Acute chest syndrome is the second most common cause of hospital admissions for patients with sickle cell disease (Rees et al., 2010). It is a pneumonia-like illness caused by infection; infarction, and fat emboli (Benton et al., 2007). Fat embolization or liquefied bone marrow fat spreads into pulmonary vessels in the body. Symptoms include bone pain, fever, chest pain, dyspnea, confusion, agitation, coma, and acute renal failure. Thirteen percent of patients with acute chest syndrome require mechanical ventilation and 3% die (Rees et al., 2010). Generally, treatment involves broad spectrum antibiotics, bronchodilators and oxygen (Rees et al., 2010). If the patient clinically deteriorates, blood transfusion is commonly recommended (Rees et al.2010). Pulmonary issues are one of the leading causes of death in patients with sickle cell disease (Claster & Vinchinsky, 2003).

Neurological Complications

Among the most debilitating effects of sickle cell disease are neurological complications, which occur in as many as one third of patients with sickle cell disease. Stroke typically occurs during the first decade of life. Neurological conditions in patients with sickle cell disease constitute a continuum of brain impairment, including overt stroke or cerebrovascular accident (CVA) and silent stroke. Cerebrovascular accident occurs in an estimated 5- 8% of children and adolescents with sickle cell disease (Ohene-

Frempong et al., 1998). Silent strokes are clinically undetectable on routine physical examination but reflect microvascular accidents that are detectable on radiography. According to the research, an estimated 20% of children and adolescents with sickle cell disease have silent infarcts (Ohene-Frempong et al., 1998). They typically involve watershed areas of the frontal lobes of the brain. By the age of 20, approximately 7-17% of children and adolescents with sickle cell anemia or Hgb SS will have sustained a stroke (Bronner et al., 1999; Ohene-Frempong et al., 1998). The mean age of stroke in these patients is 7 years with most occurring before the age of 15 (Bonner, 1999; Davies & Kingwood, 1996; Ohene-Frempong et al., 1998). This is due to occlusion of the blood vessels in the central nervous system, which may lead to cerebral vascular hemorrhaging (Bonner et al., 1999; Davies et al, 1996; Ohene-Frempong et al., 1998.). Children and adolescents with sickle cell disease who suffer a stroke are at high risk for subsequent strokes within three years following the initial incident. Lastly, stroke correlates with early death in children and adolescents with sickle cell disease (Bonner, 1999; Davies et al., 1996; Ohene-Frempong et al., 1998).

Medical Management of Sickle Cell Disease

Medical management of sickle cell disease generally focuses on the symptoms. It may consist of hydration, warmth, analgesia, prophylactic antibiotics, hydroxyurea for vaso-occlusive crises and folic acid supplementation to support red cell production. Most states have implemented newborn screening programs that have resulted in early treatment intervention thereby leading to a decrease in morbidity and mortality (Ohene-Frempong et al., 1998; Thompson & Gustafson, 1995). In general, the highest rate of

mortality occurs during the first three years of life, primarily as a result of bacterial infections. Early intervention with prophylactic penicillin has become the standard of care. Moreover, red blood transfusions are required for children who have had a stroke or are at risk for a stroke to reduce the percentage of hemoglobin S in the blood and the associated symptoms. Many symptoms of sickle cell disease can be managed through periodic outpatient clinic visits and with patient education. However, hospitalization may still be indicated.

Some promising procedures and treatments are being evaluated for the efficacy in reducing the severity of sickle cell disease. The Stroke Prevention Trial in Sickle Cell Anemia used Transcranial Doppler (TCD) ultrasound to screen patients for areas of the brain with higher than normal blood flow which is an indication of stroke. Patients deemed high risk were randomized to either chronic transfusions or standard of care (Bonner et al., 1999). Patients on chronic transfusions received regular transfusions to decrease their hemoglobin S below 30% in order to reduce their risk of stroke by 90% (Bonner et al., 1999). In another study, the Stroke with Transfusions Changing to Hydroxyurea (SWITCH), researchers compared both hydroxyurea and blood transfusions in patients with stroke (Rees et al., 2010). However, the SWITCH study ended early because of the high rate of strokes among participants in the hydroxyurea group (Rees et al., 2010). Researchers continue to examine potentially new treatment modalities such as short-chain fatty acid, membrane-active drugs, and gene therapy (Bonner et al., 1999).

Over the past decade, there have been numerous advancements in the treatment of sickle cell disease. One such advancement is hydroxyurea. Hydroxyurea is a

chemotherapeutic agent that was approved by the U.S Food and Drug Administration in 1998 as a treatment option for patients with sickle cell disease (Charache et al., 1995). Hydroxyurea works by stimulating the production of fetal hemoglobin and hematocrit. It has been shown to reduce the frequency of vaso-occlusive crises, acute chest syndrome, and consequently the need for fewer blood transfusions in patients with sickle cell disease. It has also been demonstrated to increase life expectancy for patients. In general, it is recommended to patients who experience vaso-occlusive crises and who would comply with myelosuppression monitoring. However, the long-term risks of hydroxyurea are still unknown.

Bone marrow transplantation remains the only curative treatment for sickle cell disease. The first successful bone marrow transplantation was performed 25 years ago in a child with sickle cell disease and leukemia (Buchanan, 1995). Generally, bone marrow transplantation is reserved for patients with severe disease, characterized by prior cerebrovascular accidents or recurrent vaso-occlusive crises or pulmonary complications. With strict criteria for inclusion, only a few hundred patients have been transplanted worldwide. Bone marrow transplantation is safest in patients with HLA-compatible siblings. It is estimated that fewer than 10% of children with sickle cell disease fulfill the inclusion criteria, and among this 10%, only 1 in 5 will have a matched donor to qualify for bone marrow transplantation (Davies & Roberts, 1996). However, among those who have undergone transplants, results show an overall survival rate of 92 to 94% and event-free survival of 75 to 84% at 6 and 11 years post-transplant (Bronner et al., 1999).

Although many strides have been made in the management of patients with sickle

cell disease, the life expectancy of these patients is still significantly decreased compared with the general population. For many years, sickle cell disease was a disease of early childhood (Platt et al., 1994). In 1973, the median survival age was 14.3 years, with 20% of the deaths occurring in the first 2 years of life, one third occurring before the fifth year of life, half between 5 and 30 years of age, and one sixth after the age of 30 (Platt et al., 1994). In 1994, the National Institutes of Health (NIH) sponsored the Cooperative Study of Sickle Cell Disease. Researchers found that the median survival age for patients with sickle cell anemia was 42 years for men and 48 years for women (Quinn, Rogers, & Buchanan, 2004). A recent study reported that half of all patients with sickle cell anemia only survive into their 40s. With this increase in the life span of patients with sickle cell disease, research has begun to focus on how patients are affected by, and continue to live with, factors associated with the illness.

CHAPTER 3

Models of Adaptation to Chronic Illness

Over the last twenty years, researchers have become increasingly interested in the complex relationship between stress-coping and physical as well as psychological illness. Childhood illness has been conceptualized as a potential stressor to which children and adolescents must attempt to adapt. Numerous studies show that chronically ill children and adolescents may be at risk for adjustment problems and impaired psychosocial functioning. In recent years, researchers have shifted from more descriptive accounts of children and adolescents with chronic illnesses to more conceptually and theoretically based models to understand psychosocial functioning (Casey et al., 2000). These models seek to understand the independent and combined factors that influence chronically ill children and adolescents' psychosocial functioning.

The social-ecological theory was one of the first models to examine the impact of biological and psychosocial influences on human development (Thompson & Gustafson, 1996). According to systems-theory, all levels of organization are connected to each other on a hierarchy as well as on a continuum. Change at one level affects change at all levels. The hierarchy evolves from the cell level, organ level, and to the person level, which is the highest level of the hierarchy (Thompson & Gustafson, 1996). Similarly, the person level is the lowest level on the social hierarchy, which moves from family to community. Systems-theory focuses on accommodations that are made throughout the life span between the organism and the environment (Thompson & Gustafson, 1996).

The social-ecological systems theory by Bronfenbrenner proposes a series of concentric circle that represent settings that have bidirectional influences on the child. The child is at the center and the closer concentric circles represent the family. The more distant circles represent societal values and culture. There are three major tenets of the social-ecological systems theory. They are reciprocity which means, not only does the environment affect the child but that the child affects the environment; interconnections and relations between settings, which influence the child's development and transitions through the life span (Bronfenbrenner, 1979).

According to the social-ecological systems theory, the setting most crucial to the development of the child is the family system, in particular, the parent-child interaction. The child's characteristics and behaviors influence the environment as well as the bidirectional influence between the child and parents. As a result, the child has an active role in shaping his or her environment (Thompson & Gustafson, 1996).

Systems theory and social-ecological systems theory both maintain that adaptation is an accommodation that occurs throughout the life span (Thompson & Gustafson, 1996). They are concerned with the impact of experiences on the developing child and on others in the environment and on factors that mediate and moderate developmental outcomes (Thompson & Gustafson, 1996). And lastly, both theories focus on constitutional and environmental factors and their impact on the developing child over time (Thompson & Gustafson, 1996).

The Integrated Model

The integrated model developed by Pless and Pinkerton emphasized the role of

psychological processes in adjustment to chronic illness (Thompson & Gustafson, 1996). This model proposes that adjustment is a dynamic process that continues from childhood through adulthood. Adjustment is determined by the individual's transactions with his or her environment (Thompson & Gustafson, 1996). The presence of "cybernetic circuits between family attitudes, other social factors, the child's own basic attributes and his responses to illness influence adjustment" (Pless & Pinkerton, 1975, p.30). In addition, "current functioning influences the response of others, which in turn, reciprocally influence future functioning" (Pless & Pinkerton, 1975, p. 30). Furthermore, adjustment changes over time, given that psychological functioning is based on earlier transactions (Pless & Pinkerton, 1975).

According to the integrated model, genetic, social, and family factors are determined primarily by a child's attributes. These attributes interact with facets of the disease, and with attributes of family, peers, teachers, and significant others (Pless & Pinkerton, 1975). These interactions determine the child's self-concept and coping style and, thus determine the child's level of adaptation. The child's adaptation is influenced by all these factors as well as by the way others respond to his or her condition (Pless & Pinkerston, 1975)

Life Crisis Model

In 1977, Moos and Tsu proposed the life crisis model. The life crisis model extends Pless and Pinkerton's work by incorporating psychological approaches to stress and by viewing chronic illness as a life crisis (Moos & Tsu, 1977). According to the life crisis model, the diagnosis of an illness represents a crisis and, subsequently, threatens the

person's own personal and social identity (Moos & Tsu, 1977). As a result, the person seeks to reestablish a balance by employing problem-solving strategies. If these strategies prove inadequate because of the person's coping response is insufficient, or because the event is so significant, the person will be thrown into chaos (Moos & Tsu, 1977). According to this model, the person will either come to cope successfully, resulting in personal growth or maturation, or will develop maladaptive responses, resulting in psychological impairment (Moos & Tsu, 1977).

The life crisis model emphasizes the person's cognitive appraisal of the crisis. It examines the importance of cognitive appraisal, or the individual's understanding and thoughts about his or her own illness (Moos & Tsu, 1977). Cognitive appraisal, perception of the task involved, and the selection of the coping skills are influenced by background and personal characteristics, illness-related factors, and features of the physical and sociocultural environment (Moos & Tsu, 1977). Furthermore, crisis not only affects the person with the illness but also those around him or her (Moos & Tsu, 1977).

Moos and Tsu (1977) developed seven categories of adaptive tasks related to chronic illness. Three of the tasks are illness-related and four are general life crisis. The illness-related tasks include dealing with the symptoms of the condition, and with the stress associated with treatment, and developing and maintaining an adequate relationship with the medical team (Moos & Tsu, 1977). The general adaptive tasks include perceiving a reasonable emotional balance, preserving a satisfactory self-image, maintaining a sense of competence and mastery, and readjusting goals and expectations;

preserving relationships with family and friends, and preparing for an uncertain future, in which there is the threat of loss, while maintaining hope (Moos & Tsu, 1977).

Moos and Tsu (1977) proposed several coping skills to deal with the adaptive tasks associated with illness. They include: denying or minimizing the seriousness of the situation, seeking information or social support, learning general problem-solving skills, and finding a general purpose or meaning in one's illness (Moos & Tsu, 1977). According to this model coping skills are not inherently adaptive or maladaptive, but are dependent on the illness-related situation (Moos & Tsu, 1977). Subsequently, the process of coping involves both problem-solving efforts to change the nature of the threatening situation and emotional regulation.

Stress and Coping Theory

Lazarus and Folkman's (1984) proposed their stress and coping theory to explain psychosocial adjustment. According to stress and coping theory, psychological stress is the relationship between the person and the environment. The way in which people think about the situation and themselves will influence their responses. It is influenced by appraisals or judgment, expectations or beliefs, and attributions of meaning and cause (Lazarus & Folkman, 1984). Cognitive appraisal and coping are critical mediators of stress person-environment relationships. Cognitive appraisal is the process by which the person evaluates whether a particular interaction with the environment is stressful (Lazarus & Folkman, 1984). This process involves two components which Lazarus and Folkman defined as primary and secondary appraisal (Lazarus & Folkman, 1984). Primary appraisal involves evaluating the significance of a situation in relation to

personal well-being (Lazarus & Folkman, 1984). Individuals can appraise a situation as irrelevant, benign-positive, or stressful (Lazarus & Folkman, 1984). The model proposes three types of stressful appraisal. Threat refers to a potential for harm or loss. Challenge refers to an opportunity for growth and mastery. However, both appraisal of harm and threat are associated with negative emotions and appraisals of challenge are associated with positive emotions (Lazarus & Folkman, 1984).

Personal and situational factors influence appraisals. Included among the personal factors are cognitive processes such as expectations and commitments (Lazarus & Folkman, 1984). Expectations are beliefs that can influence the way in which people perceive situations. Commitments are values and ideals that define what is important to the person. As a result, expectations and commitment clarify what is at risk in specific transactions (Lazarus & Folkman, 1984).

Secondary appraisal involves evaluating coping resources and options with regard to the situation. Coping resources include physical, social, psychological and material assets (Lazarus & Folkman, 1984). Secondary appraisal includes judgment regarding control and efficacy (Lazarus & Folkman, 1984). Appraisals of control involve evaluating the degree to which the outcome of a situation is under the individual's control, some other person's control, or a matter of chance (Lazarus & Folkman, 1984). If a situation is perceived to be under the individual's control, then the appraisal is made of personal efficacy (Lazarus & Folkman, 1984)).

Coping is the process by which the individual manages the internal and external demands of the person and environment (Folkman, Lazarus, Gruen, & DeLongis, 1986).

Once the person has appraised the situation and determined the resources available, then he or she seeks to master the situation by selecting a coping strategy or mechanism. It is generally categorized as problem-focused and emotion-focused coping (Lazarus & Folkman, 1984). Problem-focused coping refers to efforts to modify the source of the problems and emotion-focused coping refers to efforts to reduce emotional distress (Lazarus & Folkman, 1984). Stressful situations perceived as controllable elicit problem-focused coping; whereas stressful situations perceived as uncontrollable elicit emotion-focused coping. Numerous studies have shown that people use both forms of coping in almost every stressful situation. Thus, the primary goal is to optimize one's goodness of fit with the existing condition (Lazarus & Folkman, 1984).

Lazarus and Folkman hypothesized five categories of coping resources: utilitarian, including socio-economic status, money, available services; health, energy, or morale, consisting of pre-existing physical and psychiatric illness; social networks, i.e., close interpersonal relationships; general and specific beliefs, including self-efficacy, mastery and self-esteem; and problem-solving skills, which contain intellectual abilities, cognitive flexibility and analytic skills (Bradford, 1997).

Disability, Stress, and Coping Model

Varni and Wallander's (1988) disability, stress, and coping model uses Pless and Pinkerton's work on adjustment of chronically ill children and Lazarus and Folkman's work on cognitive appraisal. The model is a risk-and-resistance framework that identifies a number of risk and resistance factors that exacerbate or buffer the effects of psychosocial stress on adjustment. The model distinguishes between risk factors that are

hypothesized to increase the likelihood of adjustment problems in children and adolescents with chronic illness whereas resistance or protective factors are expected to decrease the probability of psychological dysfunction (Drotar, 2006). The model identifies three risk factors: disease and disability variables, and the child's level of functional independence; and psychosocial stressors which relate to the disease as well as other life events and daily hassles (Thompson & Gustafson, 1996). Three sets of resistance factors are also proposed. They relate to intrapersonal variables, which include temperament and problem-solving ability; social-ecological, i.e., family environment, social support, utilitarian resources; and stress processing abilities, i.e., cognitive appraisal and coping strategies (Drotar, 2006). Varni and Wallander proposed that adaptation to disease can be predicted by exploring the balance of risk and resistance factors. Resistance factors are thought to moderate the negative effects of risk factors on psychosocial adjustment and key parameters prevent maladjustment in chronically ill adolescents (Drotar, 2006).

Varni and Wallander's (1988) model places particular emphasis in the role of stressors in the person's competence. They defined stressors as the “occurrence of problematic situations requiring a solution or some decision-making process for appropriate action” (Varni & Wallander, 1988, p. 215). Coping with these stressors is the direct result of the person's competence. According to the disability, stress, and coping model competence is viewed as the effectiveness of the coping response when the person is confronted with a stressful situation (Varni & Wallander, 1988). As a result, adjustment can be understood as the level of stress experienced, which is influenced by the nature of

the problem experienced and the person's ability to cope successfully.

Numerous studies have focused on this model as it relates to chronic illness. Most studies have focused on disease parameters or severity and their influence on adaptation. The results have been somewhat mixed. One reason the results have been mixed is because severity is a difficult concept to operationally define. Consequently, most researchers have limited their studies to measuring the child's objective health and its effects on adjustment rather than exploring the parents' perception of their child's health status (Bradford, 1997).

Transactional Stress and Coping Model

The transactional stress and coping model is a theoretical model of the pathways of adaptation to a chronic illness (Hocking & Lochman, 2005). It is guided by Lazarus and Folkman's stress and coping theory and Bronfrenbrenner's ecological-systems theory. The model adapts a modified categorical approach to understanding chronic illness. Given that chronic illnesses differ in the stresses and tasks presented to the child and family in relation to age of diagnosis, type and severity of symptoms, treatment regimens, and life expectancy, it is hypothesized that there may be significant differences in psychological adjustment across illnesses (Thompson, Gustafson, Gil, Godfrey, & Murphy, 1998).

The transactional stress and coping model views chronic illness as a potential stressor to which the child and family attempt to adapt. The illness is proposed to be a function of the transactions among biomedical, developmental, and psychosocial processes (Hocking & Lochman, 2005). The model includes parameters that are illness

specific as well as common demographic and adaptation parameters that are associated across illnesses (Hocking & Lochman, 2005). Illness parameters include type, severity, and age of onset. Demographic parameters are comprised of age, gender, and socioeconomic status (Hocking & Lochman, 2005). The focus of this model is on child and maternal adaptation processes which are hypothesized to be associated with adjustment of children and their mothers more than illness and demographic parameters (Hocking & Lochman, 2005).

According to the transactional stress and coping model, adjustment to a stressor is mediated by the use of different coping processes. The first component is the cognitive processes of stress appraisal, expectations of efficacy and health locus of control, self-esteem, and causal attributions; coping methods; and social support (Thompson et al., 1988). The second component of the adaptation process is the method of coping. Adaptive coping, or problem-focused, involves changing the stressful transaction between the environment and person by attempting to change the environment or the person (Hocking & Lochman, 2005). Accordingly, illness parameters can explain some of the variance in coping and psychological adjustment in chronically ill children (Thompson et al., 1988).

The transactional stress and coping model also provides a theoretical framework for understanding the adaptation processes in both mother and children. The model proposes that only the mother's appraisal of stress has an impact on adjustment, while both the child and the mother's expectations of treatment efficacy and health locus of control have an impact on adjustment. The model also proposes that family functioning

only impacts maternal adjustment, while maternal and child adjustment affect each other reciprocally (Hocking & Lochman, 2005).

In conclusion, these conceptually based models have provided a foundation for understanding psychological adaptation in children and adolescents with chronic illness. Chronic illness is a potential stressor for child and families and thus is viewed as a life crisis. These models emphasize the significance of cognitive processes of stress appraisal, social support, and coping methods and view adaptation as a function of the goodness of fit (Thompson & Gustafson, 1996). Therefore, adaptation is a process of accommodation that occurs between the child and his or her environment. This process occurs throughout the child and adolescent's life span. And lastly, these models emphasize the critical role of parenting and the factors that influence it. Hence, chronic illness in children and adolescents presents significant challenges to the process of adaptation.

CHAPTER 4

Psychological Functioning of Children and Adolescents with Sickle Cell Disease

The psychosocial adjustment of children and adolescents with chronic illnesses has been well documented in the literature. Numerous studies have shown that chronically ill children and adolescents are at increased risk for psychosocial dysfunction. The most reported psychosocial problems cited in the literature have included poor self-concept, increased anxiety, depression, social withdrawal or aggression, and maladaptive family and peer relationships. Pless and Roghmann (1971) reviewed three relevant epidemiological studies: the United Kingdom National Survey of Health and Development, the Isle of Wight study, and the Rochester Child Health Survey. In the United Kingdom National Survey, parents, teachers, and children completed behavioral symptoms questionnaires (Pless & Roghmann, 1971). The United Kingdom National Survey showed that 25% of the children with chronic illness had two or more behavioral symptoms compared with 17% of the healthy control children (Pless & Roghmann, 1971). Similarly, 39% of children with chronic illness displayed more aggressive behavior compared with 31% of healthy children (Pless & Roghmann, 1971).

Rochester Child Health Study compared children with chronic illness with a matched control group of healthy children (Roghmann & Haggerty, 1970). The Rochester Child Health Survey revealed that children with chronic illness exhibited more abnormal behavioral symptoms than the matched control group (Roghmann & Haggerty, 1970). The United Kingdom National Survey and the Rochester Child Health Survey revealed that children with chronic illness had more school difficulties and social adjustment

problems than the healthy children (Pless & Roghmann, 1971). In addition, they were more frequently truant, more often troublesome in school, more often socially isolated, and more frequently described by their teachers as having poor attitudes toward their school work (Pless & Roghmann, 1971). In conclusion, up to 30% of children who participated in the study were impaired by psychological maladjustment (Pless & Roghmann, 1971).

The Ontario Child Healthy Study examined psychosocial functioning in 3,294 chronically ill children between the ages of 4 and 16 years from 1,869 families living in Ontario (Cadman, Boyle, Szatmari, & Offord, 1987). A mental health survey was developed that incorporated items from the Behavior Problem and Social Competence Scales of the Child Behavior Checklist (Cadman et al., 1987). The survey also included additional information to allow classification of children into psychiatric diagnostic categories from the DSM-III, including conduct disorder, attention deficit hyperactivity disorder, and neurotic disorder (Cadman et al., 1987). Parents completed the mental health survey for children between the ages of 4 and 16 years. Teacher reports were obtained for children between the ages of 4 to 11 years (Cadman et al., 1987). Lastly, child self-reports were obtained for children between the ages of 12 and 16 years (Cadman et al., 1987).

Children with a chronic illness and a disability had a risk for psychiatric disorder that was 3.4 times that of healthy children. Children with a chronic illness without disability have a risk for psychiatric disorder that was 2.1 times that of healthy children (Cadman et al., 1987). Children with at least one psychiatric disorder comprised of 31%

for children with chronic illness and disability, 22% for children with chronic illness without disability, and 14% for healthy children (Cadman et al., 1987). Neurotic disorders and attention deficit hyperactivity were the most common, especially among children with a chronic illness and a disability. Social adjustment difficulties were also more common among children with a chronic illness and a disability, while children with a chronic illness without a disability were only slightly more likely to have social adjustment problems than were healthy children (Cadman et al., 1987). Children with both a chronic illness and a disability also were at greater risk for school difficulties (Cadman et al., 1987).

In another epidemiological study examined the relation between chronic conditions and behavioral problems in children and adolescents. Parent reports of behavior problems and chronic childhood conditions were obtained from a sample of 11,699 children and adolescents aged 4 to 17 in the United States (Gortmaker, Walker, Weitzman, & Sobol, 1990). Chronic conditions were assessed using a 59-item checklist that yielded 19 chronic condition categories (Gortmaker et al., 1990). Behavior problems were assessed by the Behavior Problem Index, a brief measure adapted from the Child Behavior Checklist (Gortmaker et al., 1990). The results showed that chronic physical conditions were a significant risk factor for behavior problems (Gortmaker et al., 1990). Behavior problems about the 90 percentile were 1.55 times higher among children with a chronic illness than those without a chronic illness (Gortmaker et al., 1990). Children with a chronic illness primarily demonstrated internalizing problems and social difficulties (Gortmaker et al., 1990). Chronic health conditions were also a risk factor for

school problems (Gortmaker et al., 1990)

In a review conducted by Pless and Nolan (1991), they concluded that children and adolescents with pediatric conditions were twice as likely to demonstrate psychological maladjustment problems compared to healthy children. In another meta-analysis, Lavigne and Afire-Routman (1992) conducted a review of 87 studies. They found that children with chronic physical conditions were at increased risk for internalizing and externalizing symptoms (Lavigne & Faier-Routman, 1992). In 1994, Bennett conducted a meta-analysis of 60 studies. He concluded that children with pediatric conditions were at higher risk for depressive symptoms than physically healthy children. Similar results have been obtained from meta-analysis of the psychosocial adjustment of children and adolescents with a variety of chronic illnesses such as diabetes, cystic fibrosis, juvenile rheumatoid arthritis, and asthma.

A growing body of literature suggests that children and adolescents with sickle cell disease are at increased risk for disturbances in a range of psychosocial functions, including behavior problems, social and personal adjustment, and body image (Hurtig, Koepke, & Park, 1989). One of the earliest studies to examine the psychosocial functioning of children and adolescents with sickle cell disease was conducted by Conyard, Krishnamurthy, and Dusik (1976). They looked at a range of behaviors and found that adolescents with sickle cell disease demonstrated poor school performance, a high degree of isolation, dependence, fear of illness, and withdrawal, as well as emotional problems including poor self-image, depression, anxiety and preoccupation with death (Conyard, Krishnamurthy & Dusik, 1976). However, as with many early studies

examining the psychosocial functioning of chronically ill children and adolescents, the findings were based on clinical observations, with no standard measures of the behaviors and no comparison with test controls.

In another study, Kumar, Powars, Allen and Haywood (1976) investigated the psychological affect of sickle cell disease in school aged children. They compared 29 African American children with sickle cell disease to 26 African American children without sickle cell disease (Kumar et al., 1976). They used the General Anxiety Scale for Children, California Test, and the Piers-Harris Self-Concept Scale to measure anxiety, self-concept, personal adjustment and social adjustment (Kumar et al., 1976). The results showed that the children with sickle cell disease did not differ from the children without sickle cell disease in personal, social, and total adjustment. Similarly, the self-concept and anxiety scores were lower in children with sickle cell disease than those without sickle cell disease (Kumar et al., 1976).

In 1986, Moise examined self-esteem and adjustment problems in 33 children and adolescents with sickle cell disease. The sample included 17 males and 16 females between the ages of 8 and 16 (Moise, 1986). She used the Nowicki-Strickland Internal-External Scale, the Family Environment Scale Form R, Piers-Harris Children's Self-Concept Scale, the Stressful Life Events Checklist, California Test of Personality, and Child Behavior Profile (Moise, 1986). She found that most children with sickle cell disease had adequate self-esteem based on a range of standardized personality and adjustment measures but high levels of adjustment problems, with social adjustment particularly compromised (Moise, 1986).

Morgan and Jackson (1986) conducted one of the first studies to compare responses of mothers of adolescents with and without sickle cell disease. They compared 24 adolescents with sickle cell disease and their mothers to a matched comparison group of healthy adolescents and their mothers. They wanted to determine whether adolescents with sickle cell disease exhibited less body satisfaction, more symptoms of depression, and greater social withdrawal than healthy peers. They found that adolescents with sickle cell disease reported higher scores on the Children's Depression Inventory and on the sub-scales of the Child Behavior Checklist in depression and social withdrawal, and lower body satisfaction than healthy adolescents (Morgan & Jackson, 1986). The mothers of the children with sickle cell disease also reported higher levels of body dissatisfaction, more symptoms of depression and greater social withdrawal in the adolescents with sickle cell disease than the mothers of the healthy adolescents (Morgan & Jackson 1986). The results showed that adolescents with sickle cell disease are at higher risk for psychosocial adjustment than healthy adolescents.

In 1989, Hurtig, Koepke, and Park examined the relationship between severity of chronic illness and adjustment in children and adolescents with sickle cell disease. They recruited 70 children and adolescents from the University of Illinois at Chicago, Department of Pediatrics, Hematology Department, and Children's Memorial Hospital of Chicago (Hurtig, Koepke & Park, 1989). The children and adolescents with sickle cell disease were between the ages of 8 and 16 years (Hurtig, Koepke, & Park, 1989). They participated in a structured interview and completed the Wechsler Intelligence Scale for Children-Revised, the Piers-Harris Self-Concept Scale and the California Test of

Personality (Hurtig, Koepke, & Park, 1989). The results showed that disease takes a more serious toll on adolescents than previously reported. In addition, the study supported previous studies which suggested that the limitations imposed by sickle cell disease have a greater impact on adolescent boys than girls (Hurtig, Koepke, & Park, 1989). Physical factors, such as small size, delayed puberty, and limited physical activities are more disruptive to boys than girls (Hurtig, Koepke, & Park, 1989). In addition, younger children did better academically than adolescents (Hurtig, Koepke, & Park, 1989). Adolescent girls did better than adolescent boys, which was consistent with previous studies.

In another study conducted by Hurtig and White (1986), they investigated the degree to which the stress of chronic illness impacted adjustment in a sample of 50 children and adolescents with sickle cell disease. Various personality and social-interpersonal assessment measures were used in the study. The measures included a short form of the Wechsler Intelligence Scale for Children- Revised, the California Test of Personality, the Child Behavior Profile-Revised, Nowicki-Strickland Locus of Control Scale, and the Piers-Harris Self-Concept Scale (Hurtig & White, 1986). The results showed that children and adolescents with sickle cell disease exhibited problems in a range of adjustment variables, particularly for adolescent males and most significantly in the areas of behavior problems and social adjustment (Hurtig & White, 1986). However, methodologically, no comparison group was used in this study.

In a similar study, Lemanek, Moore, Gresham, Williamson, and Kelley (1986) examined the psychosocial adjustment of 30 children with sickle cell disease. They

compared the children with sickle cell disease to a group of 30 healthy children. The groups differed on several measures related to medical problems. The measures included the Piers-Harris Children's Scale, the Children's Depression Inventory, Behavior Rating Profile, Wechsler Intelligence Scale for Children-Revised, the Child Behavior Checklist, and the Parent Discipline Methods/ Knowledge Questionnaire. They also collected interview data which consisted of information concerning the number of household members, grade level, and average grades. The results showed that children with sickle cell disease did not differ on psychological adjustment with the healthy children. However, the children with sickle cell disease showed more behavioral problems at home and at school, as well as with peers when compared to the healthy children (Lemanek, Moore, Gresham, Williamson & Kelley, 1986). However, behavior problems were higher in both groups when their scores were compared to normative samples (Lemanek et al., 1986). This study had several major limitations. First, the researcher did not match for age, gender, and socioeconomic status. Second, the comparison group was ethnically and racially diverse which could have influenced the results.

In 1990, Seigel, Golden, Gough, Lashley, and Sacker assessed depression, self-esteem, and life events in adolescents with chronic diseases. The sample consisted of 20 adolescents with sickle cell disease, 40 adolescents with asthma, and 20 adolescents with diabetes. The control group consisted of 100 healthy adolescents. Seigel and his colleagues matched for age and socioeconomic status (Seigel et al., 1990). Both groups of adolescents were administered the Beck Depression Inventory, Rosenberg Scale of Self-Esteem, and the McCutcheon Life Events Checklist (Seigel et al., 1990). The results

showed that adolescents with chronic disease had higher depression and lower self-esteem scores than the adolescents in the control group; however, the life event score did not differ between the two groups (Seigel et al., 1990). They also showed that depression, self-esteem and life event did not differ among the three disease groups.

A study conducted by Brown, Kaslow, Doepke, Buchanan, Eckman, Baldwin, Goonan, and Schoenherr (1993) compared adolescents with sickle cell disease to healthy siblings. They looked specifically at depression and somatic complaints. They found that adolescents with sickle cell disease exhibited more depressive symptoms than their healthy siblings (Brown, Kaslow, Doepke, Buchanan, Eckman, et al., 1993). Additionally, they were perceived by their mothers as having more internalized behaviors and higher levels of somatic complaints than their healthy siblings (Brown et al., 1993).

In 1993, Thompson, Gil, Burbach, Keith, and Kinney (1993) examined the psychological adjustment of children with sickle cell disease. Ninety-one children between the ages of 7 and 17 and their mothers were recruited from the Duke University Comprehensive Sickle Cell Center (Thompson, Gil, Burbach, Keith, & Kinney, 1993). Mothers completed the Symptom Checklist 90-Revised and the Missouri Children's Behavior Checklist (Thompson et al., 1993). Children completed the Coping Strategies Questionnaire and the Child Assessment Schedule (Thompson, Gil, Burbach, Keith, & Kinney, 1993). Six-four percent of the children with sickle cell disease were perceived by their mothers to have a behavior problem, and 50% of the children reported symptoms that met the criteria for a *DSM-III* diagnosis (Thompson et al., 1993). Internal behavior problems and anxiety were the most frequent types of adjustment problems (Thompson,

et al., 1993). In addition, children with mother-reported behavior problems had mothers with significantly higher anxiety and depression compared with mothers of children without reported problems (Thompson et al., 1993). As a result, children with sickle cell disease were perceived by their mothers to be at risk for psychosocial dysfunction (Thompson et al., 1993).

Barbarin, Whitten, and Bonds (1994) investigated psychosocial problems in urban and poor children with sickle cell disease. They recruited a sample of 327 children with sickle cell disease and their parents. Participants completed a structured interview during their routine clinic appointment (Barbarin et al., 1994). Parents participated in a psychosocial interview that included questions about psychosocial functioning and academic achievement (Barbarin et al., 1994). They found that children with sickle cell disease were more prone to anger, hopelessness, depression, and shame (Barbarin et al., 1994). They also reported more pain and were more likely to be overprotected by their parents than their siblings (Barbarin et al., 1994). Lastly, older children with sickle cell disease reported a higher rate of school failure than younger children with sickle cell disease (Barbarin et al., 1994).

Thompson, Gil, Keith, Gustafson, George, and Kinney (1994) examined psychosocial adjustment over a 10-month period in children with sickle cell disease and their parents. They recruited 30 children between the ages of 7 and 14 years and their mothers from Duke University Medical Center. Children completed the Children's Health Locus of Control Scales, the Self-Worth sub-scale, the Self-Perception Profile for Children and the Coping Strategies Questionnaire. Mothers responded to the Symptoms

Checklist 90-Revised, the Child Assessment Schedule, and the Missouri Children's Behavior Checklist (Thompson et al., 1994). The results showed that the poor rate of psychological adjustment in children with sickle cell disease remained relatively stable over time (Thompson et al., 1994). In addition, these researchers also found less stability in child adjustment by child report than by mother report (Thompson et al., 1994). However, a major limitation of this study is the small sample size. Although, this study was one of the first studies to examine psychosocial functioning over time, the small sample size limits the studies' generalizability.

Yang, Cepeda Price, Shah, and Mankad (1994) examined depression, anxiety, and behavioral problems in 38 children and adolescents between the ages of 6 and 18 with sickle cell disease and a demographically matched control group of adolescents without sickle cell disease. They matched for age and race. Each child and adolescent completed the Children's Depression Rating Scale Revised and participated in a structured interview with a child psychiatrist (Yang et al., 1994). They found that 13% of the children and adolescents exhibited depressive symptoms, 10% reported anxiety, and 8% behavioral problems (Yang et al., 1994). However, the structured interview revealed that the prevalence of clinical depression in children and adolescents with sickle cell disease was not higher compared with that of the children without sickle cell disease (Yang et al., 1994). Although children and adolescents tended to report higher somatic complaints, worries about death, and self-esteem problems compared to healthy children and adolescents, these symptoms did not necessarily meet criteria for a psychiatric disorder.

In a similar study, Lee, Phoenix, Brown, and Jackson (1997) looked at depression

and social competence in children with sickle cell disease. They compared 14 children with sickle cell disease with 14 healthy siblings. Each child completed the Depression Self Rating Scale (Lee et al., 1997). They found that the children with sickle cell disease scored lower on measures of perceived competence but the healthy siblings scored higher on measures of depression (Lee et al., 1997).

Thompson, Gustafson, Gil, Godfrey, and Murphy (1998) looked at specific patterns of psychological adjustment and cognitive adaptation in children with cystic fibrosis and sickle cell disease. They recruited 43 children between 7 and 12 years of age from the Duke University Medical Center Cystic Fibrosis Clinic and 49 children between 7 and 12 years of age from the Duke University Medical Center Comprehensive Sickle Cell Program. They matched on maternal education, socioeconomic status, and gender. Severity of cystic fibrosis was assessed with the Shwachman Clinic Evaluation System and severity of sickle cell disease was assessed based on phenotype, number of sickle cell disease complications, and frequency of pain crises (Thompson et al., 1998). The children completed the Self-Perception Profile, the Child Assessment Schedule, and the Children's Health Locus of Control Scale (Thompson et al., 1998). The results showed that anxiety disorders were most prominent for both groups; children with cystic fibrosis had significantly higher levels of externalizing symptoms and higher frequency of oppositional disorder and conduct disorder than children with sickle cell disease (Thompson et al., 1998). They concluded that fostering a sense of efficacy and internal health locus of control may be instrumental in promoting adaptation in children with cystic fibrosis and enhancing feelings of self-worth may be a salient intervention in

children with sickle cell disease (Thompson et al., 1998).

In a follow-up study, Thompson, Gustafson, Gil, Kinney and Spock (1999) examined the adjustment of children and adolescents with sickle cell disease and cystic fibrosis and their mothers. The study was comprised of 59 children with cystic fibrosis and 50 children with sickle cell disease and their mothers. The participants were recruited from the Cystic Fibrosis Center and Sickle Cell Disease Center at Duke University Medical Center. The purpose of the study was to investigate the psychological adjustment of children and adolescents 2-years after the initial assessment. Severity of cystic fibrosis was determined by the Shwachman Clinical Evaluation System and severity of sickle cell disease was determined by genotype, total number of sickle cell related complications and frequency of pain crises (Thompson et al., 1999). Psychosocial adjustment in children with cystic fibrosis was assessed using the Children's Health Locus of Control Scale and the Self-Worth sub-scale of the Self-Perception Profile. Similarly, psychosocial adjustment in children with sickle cell disease as assessed using the Coping Strategies Questionnaire. Mothers completed the Ways of Coping Questionnaire, the Symptoms Checklist 90-Revised and the Missouri Children's Behavior Checklist. Findings from the study showed stability over time in adjustment and behavior patterns in children with sickle cell disease and cystic fibrosis (Thompson et al., 1999). In addition, the findings showed that illness-related stress was associated with family conflict and poor adjustment in children with sickle cell disease (Thompson et al., 1999).

Casey, Brown, and Bakeman (2000) conducted a study that assessed the impact of

risk and resistance factors on the psychosocial adjustment of children and adolescents with sickle cell disease. They recruited 118 children and adolescents with sickle cell disease and their caregivers from a comprehensive National Institutes of Health sickle cell center that serves primarily low-income families. Subjects completed the Vineland Adaptive Behavior Scales, the Coping Strategies Inventory, the Child Behavior Checklist, and the Family Adaptability and Cohesive Environmental Scales II (Casey et al., 2000). The study found that adaptive behavior was associated with child maladjustment, severity of disability was associated with disability stress, child competence was associated with child maladjustment and adaptive behavior and stress did not mediate the association between severity of disability and maladjustment (Casey et al., 2000). A major limitation of this study is its generalizability to the larger population. Whether the data can be attributed to stressors associated with a chronic condition or to other factors cannot be determined from this study. In addition, previous studies have suggested that the existing standardized measurements of adaptability and cohesion may not be relevant to African Americans (Casey et al., 2000).

In 2000, Burlew, Telfair, Colangelo, and Wright examined the relationship between psychosocial factors and biomedical risk factors in predicting adolescent adaptation to sickle cell disease. They recruited 90 African American adolescents between the ages of 14 and 19 with sickle cell disease and their parents from a multi-site Cooperative Study of Sickle Cell Disease. Adolescents completed the State-Trait Anxiety Inventory, Beck Depression Inventory, the Rathus Assertiveness Schedule, the Rosenberg Self-Esteem and parents completed the Family Environment Scale, the Coping Health

Inventory for Parents and the Family Relations Index (Burlew et al., 2000). These researchers found that intrapersonal, stress-processing and social ecological factors were significant predictors of adaptation; however, biomedical factors did not predict adaptation (Burlew et al., 2000). There was no evidence that psychosocial factors moderated the relationship between biomedical risk factors and adaptation. In conclusion, psychosocial factors were better predictors of adaptation than biomedical risk factors (Burlew et al., 2000).

In 2001, Key, Brown, Marsh, Spratt, and Recknor looked at depressive symptoms in adolescents with various chronic illnesses. Using the Beck Depression Inventory, they compared 125 adolescents with various chronic illnesses, including cystic fibrosis, diabetes, spina bifida, sickle cell disease and asthma to 21 healthy adolescents. They found that adolescents with a chronic illness reported symptoms of moderate to severe depression. In addition, the frequency of depression was highest in adolescents with sickle cell disease (Key et al., 2001).

Trzepacz, Vannatta, Gerhardt, Ramey, and Noll (2004) researched emotional, social, and behavioral functioning in children with sickle cell disease and comparison peers from the perspective of primary and secondary caregivers. The study consisted of 70 children with sickle cell disease and 67 comparison peers (Benton et al. 2007; Trzepacz et al., 2004). The parents completed the Child Behavior Checklist during home visits. The children with sickle cell disease were perceived by their caregivers to have more internalizing behaviors and more externalizing problems than the comparison peers (Trzepacz et al., 2004). Their caregivers also perceived them as being less successful with

social interactions and academic performance than comparison peers (Trzepacz et al., 2004). However, this study has several limitations. The researchers collected data about the emotional, social, and behavioral functioning of children with sickle cell disease and peers with one measurement, and the sample size was also relatively small and lacked sufficient power to examine the effects of gender and age.

Lastly, Simon, Barakat, Patterson, and Dampier (2009) investigated symptoms of depression and anxiety in adolescents with sickle cell disease. They hypothesized that adolescents with sickle cell disease will report higher levels of depression and anxiety as compared to their healthy siblings. They recruited 44 adolescents with sickle cell disease and 15 healthy siblings from an East Coast comprehensive sickle cell center. Interestingly, they found that the adolescents with sickle cell disease and the healthy siblings both reported low levels of depression and anxiety as measured by the Behavioral Assessment System for Children (Simon et al., 2009). The adolescents with sickle cell disease also reported high levels of self-esteem and low levels of inadequacy (Simon et al., 2009). Furthermore, adolescents with sickle cell disease and their healthy siblings did not differ significantly.

Conclusion

With some notable exceptions, most research demonstrated that adolescents with sickle cell disease have a wide range of adjustment problems and an elevated risk for adjustment problems when compared to healthy controls. Adolescents with sickle cell disease have less body satisfaction, less academic success, less social and nonsocial activity, and a poorer self-concept than healthy controls. A meta-analysis investigating

adolescents with different chronic medical problems revealed that adolescents with sickle cell disease had higher rates of psychosocial dysfunction than control groups (Hocking & Lochman, 2005). Researchers have found that adolescents with sickle cell disease may be at increased risk for social adjustment problems than children without sickle cell disease due to disease symptoms, physical disabilities and treatment modalities, such as medication, physical therapy, and surgery which place a heavy strain on their capacity for effective coping. Cognitive, emotional, behavioral, and social problems commonly occur. The literature shows that children and adolescents with sickle cell disease are more at risk for psychiatric problems, social isolation and school performance than are healthy adolescents of the same age. Some studies of adolescents with sickle cell disease have reported that the rates of behavior problem range from 3—64%. Other studies have reported that 42.5-50% of adolescents with sickle cell disease qualify for a DSM-III diagnosis; with an anxiety disorder being the most frequent (Hocking & Lochman, 2005).

While many studies have examined the psychosocial adjustment of children and adolescents with sickle cell disease, they are not without their limitations and weaknesses. One of the major limitations of the research done in this area is many of these studies failed to incorporate reports from children with sickle cell disease and their mothers and relied on reports from physicians, which makes generalizability of the findings questionable. In addition, many of these studies were conducted more than 20-years ago and are outdated. Another major weakness of the research done in this area is most studies used small samples due to difficulties in identifying and recruiting participants, which diminishes the weight of these studies. And lastly, comparison groups

with similar demographic characteristics were not included in many of the studies with an overwhelming number of studies not matching for age and socioeconomic status.

This present study will employ a case-control design to compare the psychosocial functioning and academic achievement in siblings with and without sickle cell disease. This will improve on previous research by using a design that has higher internal validity. This study will also use convenience sampling to recruit (N=150) 100 siblings with and without sickle cell disease and their 50 caregivers from the Children's National Medical Center Sickle Cell Program in Washington, DC. While the aforementioned studies show that children and adolescents with sickle cell disease are at risk for psychosocial dysfunction, many of these studies used small sample sizes and inappropriate comparison groups. This study will use siblings without sickle cell disease for the comparison group and will physically control within the design for age, socioeconomic status, family environment and parent education. Furthermore, this study will examine academic achievement in this particular population which few studies have done. And lastly, this researcher will use measures that have been used in previous studies to assess psychosocial functioning in African American children and adolescents. This study hypothesis is that psychosocial functioning and academic achievement will be lower in siblings with sickle cell disease than in siblings without sickle cell disease.

CHAPTER 5

Methods

A substantial body of literature provides strong evidence that chronically ill adolescents are at risk for psychosocial dysfunction. However, few current studies have clearly demonstrated that adolescents with sickle cell disease are at risk for psychosocial dysfunction and problems with academic achievement. The aim of the study was to compare the psychosocial functioning and academic achievement in siblings with and without sickle cell disease. This study was designed to build on prior research conducted at the Duke University Medical Center, Children's Hospital of Philadelphia, and St. Christopher's Hospital for Children in Philadelphia by using multiple measures to examine the psychosocial functioning and academic achievement in a substantial sample of siblings with and without sickle cell disease. This study design physically controlled for age, socioeconomic status, family environment, and parent education between sickle cell subjects and comparisons. In addition, this researcher used measures that had not been used to assess psychosocial functioning in African American adolescents in previous studies as well as a demographically matched comparison group. Data were collected from both siblings and their caregivers.

Design

This study employed a case control design to compare the psychosocial functioning and academic achievement in siblings with and without sickle cell disease. A case-control design was used for this study to increase internal validity over other correlational studies. The cases were siblings with sickle cell disease and their controls

were siblings without sickle cell disease within the same age range. The dependent variable was sickle cell disease or not and the independent variables were psychosocial functioning, academic achievement, degree of depression, anxiety, aggression, rule-breaking behavior, oppositional defiant behavior, conduct disorder, social problems, somatic complaints, attention deficit hyperactivity disorder, sluggish cognitive temperament, posttraumatic stress disorder and obsessive compulsive disorder. This researcher physically controlled for age, family environment, parent education, and socioeconomic status through the use of paired siblings with and without sickle cell disease from the same household. This study used convenience sampling to recruit sibling with and without sickle cell disease and their caregivers for the study. One of the biggest criticisms of convenience sampling is sampling bias but our recruitment site and methods resulted in greater representation of the population.

Setting

The Children's National Medical Center Sickle Cell Program is the largest provider of sickle cell services in the Washington, DC metropolitan area and the second largest pediatric sickle cell program in the United States. Hematologists, fellows, residents, medical students, nurse practitioners, registered nurses, social workers, psychologists, and allied health care professionals follow 1,200 patients from birth to age 21 with sickle cell disease; of these 359 are between the ages of 12 and 18 years. Health care providers strive to meet the medical, social, and emotional needs of patients and their families through disease education, health maintenance, preventative therapies, and clinical trials.

Sample Population and Sample Size

This researcher used convenience sampling to recruit subjects (N=133) 88 adolescents, ages 12 to 18, with and without sickle cell disease and 45 primary caregivers from the Children's National Medical Center Sickle Cell Program in Washington, DC. The sample was composed of all African American siblings and their primary caregivers. The mean age for siblings with sickle cell disease was 14.93 (SD=1.97) and the mean age for siblings without sickle cell disease was 15.2 (SD=2.0). Fifty-eight percent of females and 42% of males with sickle cell disease completed the study. The most common hemoglobinapathy was hemoglobin SS. Thirty-seven percent of females and 63% of males without sickle cell disease completed the study. Ninety-six percent of female primary caregivers; 4% of male primary caregivers completed the study. Fifty-six percent of primary caregivers were married. Sixty-seven percent of caregivers were employed. Seventy-one percent of primary caregivers had some college or vocational training. Twenty-two percent of families reported incomes between $40,000-49,999. Lastly, 73% of families lived in Maryland. Table 1 presents results from the sociodemographic questionnaire.

Table 1 Sociodemographic Information

	N	*(%)*	*Mean*
Adolescents with SCD			
Age	45		14.93
Gender			
Male	19	42%	
Female	26	58%	
SCD type			
Hb SS	29	64%	
Hb SC	13	29%	
Hb Beta	3	7%	
Adolescents without SCD			
Age	43		15.2
Gender			
Male	27	63%	
Female	16	27%	
Primary Caregivers			
Gender	45		
Male	2	4%	
Female	43	96%	
Marital Status			
Married	25	56%	
Single	11	24%	
Divorced	6	13%	
Widowed	3	7%	
Employment Status			
Employed	30	67%	
Unemployed	9	20%	
Disabled	3	7%	
Retired	3	6%	
Education			
High school diploma/ GED	10	22%	
College/vocational training	32	71%	
Some high school	3	7%	
Income			
< $25, 999	6	13%	
$26,000-39,999	5	11%	
$40,000-49,999	10	22%	
$50,000-74,999	10	22%	
$75,000-99,999	7	16%	
$100,000-129,999	5	11%	
>$130,000	2	4%	
Geographic Location			
Maryland	33	73%	
District of Columbia	12	27%	

Adolescent Eligibility Criteria

Eligibility inclusion criteria included:

-Adolescents between the ages of 12 and 18 diagnosed with hemoglobin SS, SC, and sickle cell β thalassemia, which includes sickle cell ß+ thalassemia and sickle cell ß° thalassemia (Hb SB O thal)

-Sibling of the subject child with sickle cell disease also between the ages of 12 and 18 without sickle cell disease.

-Voluntary consent from primary caregivers for siblings who were between the ages of 12 and 18 with and without sickle cell disease

-Assent from siblings with and without sickle cell disease who were between the ages of 12 and 17

-Subject child with sickle cell disease and his/her sibling had to reside in the same household.

-By "primary caregiver" this researcher meant biological parent or Legally Authorized Representative (LAR). By "sibling" this researcher meant full or half sibling.

-Sibling with and without sickle cell disease and their primary caregiver had to be able read and speak English.

Recruitment Procedures

This researcher approached children, ages 12 to 18, with sickle cell disease and their primary caregiver about the study during clinic visits and inpatient hospitalizations. If the child with sickle cell disease had a full or half sibling without sickle cell disease within the same age range, this researcher described the purpose of the study as well as

risks and benefits of participating in the study. This researcher explained that they could decide not to take part in the study without any penalty or loss of benefits. This researcher then gave them time to consider the study and ask questions. If the child with sickle cell disease and his/her sibling without sickle cell disease and their primary caregiver agreed to participate, they signed the Children's National Medical Center/ University of Pennsylvania consent form and/ or assent form, if appropriate. They then received instructions for completing the measures on SurveyMonkey. Upon completion of the study, a $25 gift card was given to the child and his/her sibling and their caregiver.

If the sibling without sickle cell disease was not present during the clinic visit or hospitalization, this researcher obtained consent from the caregiver and sibling with sickle cell disease. This researcher then obtained verbal permission from the primary caregiver to call and e-mail the sibling without sickle cell disease, in order to explain the purpose of the study. If he/ she agreed to participate, this researcher provided instructions for completing the Children's National Medical Center/ University of Pennsylvania assent or consent form and the measure on SurveyMonkey. Follow-up telephone calls were made to answer any additional questions the siblings and their caregiver had about the study. If the caregiver and/or child with sickle cell disease or his/her sibling without sickle cell disease did not agree to participate in the study, recruitment immediately ended, and they were thanked for their time.

A total of 133 siblings with and without sickle cell disease and primary caregivers were recruited over an eight month period (January 2012-September 2012) from the Children's National Medical Center Sickle Cell Program. This researcher approached 62

adolescents with sickle cell disease. Forty-five siblings with sickle cell disease completed the study. Forty-three siblings without sickle cell disease completed the study. Forty-five primary caregivers completed the study. Two siblings without sickle cell disease did no complete the study.

Retention, Subject Payment, Tracking Procedures

Each sibling with and without sickle cell disease and their caregiver were compensated with a $25 gift card, a $5 meal ticket, and a $5 parking sticker. This was consistent with the norms at Children's National Medical Center for subject participation payment.

Data on Refusers and Drop-outs

The only data collected from siblings with and without sickle cell disease and caregivers who refused to participate in the study were observer estimates of age, sex, and ethnicity. Fifteen primary caregivers and siblings with sickle cell disease refused to participate in the study. Eleven primary caregivers refused to participate in the study because of time constraints. Four siblings with sickle cell disease did not have a full or half-sibling without sickle cell disease between the ages of 12 and 18.

Measures

Adolescent Measures

The Youth Self Report was administered to both the child with sickle cell disease and his/her sibling to measure internalizing behavior, withdrawal/ depression, anxiety/depression, affective problems, depression, anxiety, social problems, social competence, externalizing behavior, aggressive behavior, rule-breaking behavior, conduct disorder, oppositional defiant disorder, obsessive compulsive disorder, posttraumatic

disorder, somatic complaints, somatic problems attention deficit hyperactivity disorder, thought problems, attention problems, and academic achievement. The YSR is 112 Likert scale that is used to measure emotional and behavioral functioning in children and adolescents between the ages of 11 and 18. Items are rated on a three-point scale ranging from 0 (“Not True”) to 2 (“Very True or Often True”) (Schwartz, Radcliffe, & Barakat, 2009). A majority of the items on the Youth Self Report correspond to those on the Child Behavior Checklist. The Youth Self Report has adequate psychometric properties. Test-retest reliability ranges .47 to .79. The range for internal consistency is .71 to .95 (Achenbach, 1991). Multiple studies have demonstrated acceptable criterion and construct validity (Achenbach, 1991). The alpha coefficient on the Youth Self- Report is above .80 (Achenbach & Rescorla, 2001)

Parent Measures

The Child Behavior Checklist was completed by the primary caregiver for both the child with sickle cell disease and his/her sibling. This measure parallels the Youth Self Report and measured the same concepts but from the parents' perspective. It is a widely used instrument in both clinical settings because of its positive psychometric properties (Casey et al., 2000; Lutz et al., 2004). The CBCL is a 118 item parent-report questionnaire that describes specific behavioral and emotional problems (Casey et al., 2000; Lutz et al., 2004). It assesses internalizing (anxious and depressive) and externalizing (aggressive, hyperactive, and noncompliant) behaviors. It also measures social withdrawal, somatic complaints, and anxiety (Casey et al., 2000; Lutz et al., 2004). And lastly, the CBCL measures academic achievement. Caregivers rate their child for how true each item is

within the past 6 months using the following scale: 0 = not true; 1 = somewhat or sometime true; 2 = very true or often true. The individual items of this questionnaire cluster into the following clinical scales: Anxious/Depressed, Withdrawn/Depressed, Somatic Complaints, Social Problems, Thought Problems, Attention Problems, Rule-Breaking Behavior, and Aggressive Behavior. Test-retest ranges from 0.61 to 0.96 across scales (Achenbach & Edelbrock, 1984). The alpha coefficient is above .80 (Achenbach & Rescorla, 2001). The CBCL demonstrates sound content, construct, and criterion related validity on subscales.

Cronbach's alpha coefficient was conducted to assess for internal consistency with the current sample of siblings with and without sickle cell disease and their primary caregivers. The Cronbach's alpha coefficients for the Youth Self-Report was χ=0.84, and the Child Behavior Checklist was χ =0.73, indicating good internal consistency.

•*Demographic Questionnaire*

Demographic information collected included marital status, occupation, gross family income, home address, and caregiver's highest educational level.

Analysis

The Statistical Package for the Social Sciences (SPSS + PC/ Windows version) was used to organize, manage, and analyze the data. Descriptive statistics were used to generate means and standard deviations on all interval measures and percentages for categorical variables Paired t-tests were used to assess differences in psychosocial functioning and academic achievement in siblings with and without sickle cell disease and to test the hypotheses that psychosocial functioning and academic achievement would lower in

siblings with sickle cell disease than in siblings without sickle cell disease.

Confidentiality

Protection against breaches of confidentiality: This research met with children with and without sickle cell disease and their caregivers in consultation rooms or inpatient rooms.

Facilities and Equipment

Children's National Medical Center is a 279 bed pediatric hospital with 3 intensive care units and 240 full time staff that provides general community and regional tertiary care. Children's National Medical Center operates several regional satellite centers in Virginia, the District of Columbia, and Maryland as well as an organized network of primary care with local pediatricians. The referral area is approximately 4.5 million people. Children's National Medical Center staff is trained in the management and execution of research protocols involving adolescents, subject monitoring, and data collection.

Computer: Faculty and staff utilize high performance PC-based computing systems. Software includes Microsoft Office. Access to the Internet and electronic bibliographic searches are provided as well.

Office: This researcher used office space located in the main hospital. This researcher also used conference rooms and consultation rooms in the Center for Cancer and Blood Disorders.

Human Subjects (Risks & Benefits)

This researcher was especially sensitive to the distress of families and adolescents living with a chronic medical illness. She was cognizant of my relationship with patients and families. As a result, she explained my role as a researcher to them. She did not use her

relationship in any way to coerce patients and families into consenting for the study. She also obtained approval from the IRBs at the University of Pennsylvania and Children's National Medical Center. All adolescents were asked to assent if under 18 years and to consent if 18 years old. In addition, primary caregivers were asked to consent to the study if the child with sickle cell disease and his/her sibling were under the age of 18. Lastly, if the child was 18 years old, the caregiver gave consent for his/her own participation in the study.

Adequacy of Protection against Risks

Emergency Procedures: Any distressed adolescents were referred to the psychiatric liaison on-call or to the emergency department. This information was available in our consent and assent forms.

Risks and Side Effects

The level of risk was primarily that of emotional arousal related to chronic illness. This researcher was primarily responsible for safety. There were no physical risks associated with the study procedure, and no referrals were made to the psychiatric liaison or emergency department.

CHAPTER 6

Results

This study tested the hypotheses that psychosocial functioning and academic achievement will be lower in siblings with sickle cell disease than in siblings without sickle cell disease. Paired t-tests were conducted to compare psychosocial functioning and academic achievement in siblings with and without sickle cell disease. Table 2 presents the results for measures of internalizing behaviors in siblings with and without sickle cell disease. Siblings with sickle cell disease (M=56.84, SD=6.32) and without sickle cell disease (M=52.40, SD=3.74), t=4.52, p=.00 reported clinically significant results for withdrawal/depression. Similarly, they (M=53.53, SD=4.74) reported statistically significant results for posttraumatic stress disorder as compared to their healthy siblings (M=51.84, SD=3.80), t=2.00, p=.05. They reported similar results for somatic complaints and somatic problem. Lastly, siblings with and without sickle cell disease reported clinically significant results for affective problems and anxiety.. Results for measures of externalizing behaviors (Table 3) did not differ significantly between siblings with and without sickle cell disease. These results provide support for the hypotheses that siblings with sickle cell disease are at increased risk for poor psychological functioning.

Table 2

Internalizing Behaviors in Siblings with and without Sickle Cell Disease

	Siblings with SCD		Siblings without SCD		*t*	*p*
	M	*SD*	*M*	*SD*		
Withdrawn/Depressed	56.84	6.32	52.40	3.74	4.33	.00*
Anxious/ Depressed	54.21	4.95	53.53	4.91	.64	.52
Affective Problems	54.63	5.88	52.86	4.14	1.84	.07**
Anxiety Problems	53.65	4.66	52.16	3.55	1.75	.09**
Somatic Complaints	56.74	7.73	51.74	3.46	4.01	.00*
Somatic Problems	57.23	7.39	52.63	4.04	3.67	.00*
Obsessive Compulsive	53.02	4.38	52.30	4.00	.81	.42
PTSD	53.53	4.74	51.84	3.80	2.00	.05*
Total Internalizing Behavior	53.70	9.01	46.51	7.73	4.52	.00*

Note: *p<.05, **p<.10

Table 3

Externalizing Behaviors in Siblings with and without Sickle Cell Disease

	Siblings with SCD		Siblings without SCD		*t*	*p*
	M	*SD*	*M*	*SD*		
Rule Breaking Behavior	54.19	6.86	54.58	5.93	-.32	.75
Conduct Problems	53.95	6.21	53.63	5.50	.31	.76
Aggressive Behavior	52.44	5.84	51.44	3.55	.93	.36
Oppositional Defiant	52.37	3.95	51.93	3.61	.53	.60
Total Externalizing Behavior	46.88	10.35	47.33	8.82	-.22	.83

Note: *p<.05

Table 4 presents results for social competence in siblings with and without sickle cell disease. Siblings with sickle cell disease (M=43.52, SD=9.55) reported clinically significant results for social competence compared to their healthy siblings (M=50.38,

SD=7.23), t=-3.78, p=.00. They reported similar results for social problems. Results for social problems are also presented in Table 4.

Table 4

Social Competence/ Interpersonal Functioning in Siblings with and without Sickle Cell Disease

	Siblings with SCD		**Siblings without SCD**		***t***	***p***
	M	***SD***	***M***	***SD***		
Social Problems	57.23	7.39	52.63	4.04	3.67	.00*
Social Competence	56.74	7.73	51.74	3.46	4.01	.00*

Note: *p<.05

Table 5 presents results for academic achievement in siblings with and without sickle cell disease. Siblings with sickle cell disease (M=52.47, SD=3.71) and siblings without sickle cell disease (M=51.19, SD=2.07), t=2.10, p=.04 reported statistically significant results for thought disorders. However, siblings with sickle cell disease (M=2.29, SD=.55) and siblings without sickle cell disease (M=2.27, SD=.53), t=.22, p=.83 reported statistically insignificant results for academic achievement. For attention deficit hyperactivity disorder, siblings with (M=53.35, SD=5.22) and without sickle cell disease (M=52.77, SD=4.19), t=.61, p=.55 reported clinically insignificant results. Siblings with sickle cell disease (M=53.07, SD=4.30) and without sickle cell disease (M=52.23, SD=3.34), t=.96, p=.34 reported similar results for attention problems.

Table 5

Academic Achievement in Siblings with and without Sickle Cell Disease

	Siblings with SCD		**Siblings without SCD**		***t***	***p***
	M	***SD***	***M***	***SD***		
Thought Problems	52.47	3.71	51.19	2.07	2.10	.04*
Attention Problems	53.07	4.30	52.23	3.34	.96	.34
ADHD	53.35	5.22	52.77	4.19	.61	.55
Academic Achievement	2.29	.55	2.27	.53	.22	.83

Note: *p<.05

Table 6 presents parent-reported results of children with and without sickle cell disease on measures of internalizing behaviors. Children with sickle cell disease (M=54.96, SD=8.45) were perceived by their parents to be at increased risk for internalizing behaviors compared to their healthy siblings (M=47.31, SD=8.16), t=4.83, p=.00. Similarly, they (M=56.49, SD=6.43) were perceived by their parents to be at increased risk for withdrawal/depression than their healthy siblings (M=53.07, SD=3.52), t=3.31, p=.00. They were also perceived by their parents to be at increased risk for affective problems, somatic complaints, and somatic problems. At the same time, however, children with sickle cell disease were not perceived by their parents to be at increased risk for anxiety and other related disorders, including obsessive -compulsive disorder and posttraumatic stress disorder. Their parents reported similar results for externalizing behaviors. These results are presented in Table 7. Based on these results, children with sickle cell disease were perceived by their parents to be at increased risk for internalizing behaviors.

Table 6

Primary Caregiver: Internalizing Behaviors in Children with and without Sickle Cell Disease

	Children with SCD		Children without SCD		*t*	*p*
	M	*SD*	*M*	*SD*		
Withdrawn/Depressed	56.49	6.43	53.07	3.52	3.31	.00*
Anxious/ Depressed	53.42	4.73	52.22	3.84	1.55	..13
Affective Problems	56.93	6.90	52.91	3.76	3.61	.00*
Anxiety Problems	55.47	5.53	53.93	4.95	1.39	.17
Somatic Complaints	60.76	6.82	53.47	5.28	5.34	.00*
Somatic Problems	60.67	7.40	53.13	5.34	5.36	.00*
Obsessive Compulsive	51.53	4.08	52.53	4.36	-1.39	.17
PTSD	52.73	4.51	52.2	4.02	.66	.51
Total Internalizing Behavior	54.96	8.45	47.31	8.16	4.83	.00*

Note: *p<.05

Table 7

Primary Caregivers: Externalizing Behaviors in Children with and without Sickle Cell Disease

	Children with SCD		Children without SCD		*t*	*p*
	M	*SD*	*M*	*SD*		
Rule Breaking Behavior	54.29	6.54	54.78	6.26	-.38	.70
Conduct Problems	53.44	6.01	52.64	4.67	.78	.44
Aggressive Behavior	53.89	7.01	52.67	4.22	.96	.34
Oppositional Defiant	52.89	5.07	52.09	3.84	.83	.41
Total Externalizing Behavior	51.69	7.96	50.07	7.16	1.07	.29

Note:*p<.05

Table 8 presents parent-reported results for social competence. Children without sickle cell disease (M=52.11, SD=3.96) were perceived by their primary caregivers to be at increased risk for poor social competence compared to their siblings with sickle cell disease (M=42.2, SD= 13.75), t= -4.71, p=.00. Parents reported insignificant results for social problems.

Table 8

Primary Caregivers: Social Competence/ Interpersonal Functioning in Children with and without Sickle Cell Disease

	Children with SCD		**Children without SCD**		***t***	***p***
	M	***SD***	***M***	***SD***		
Social Problems	54.64	4.92	53.20	4.78	1.30	.20
Social Competence	42.2	13.75	52.11	3.96	-4.71	.00*

Note: *p<.05

Table 9 presents parent-reported results for academic achievement in children with and without sickle cell disease. Children with sickle cell disease (M=54.91, SD=5.49) were perceived by their parents to be at increased risk for attention deficit hyperactivity disorder than their siblings without sickle cell disease (M=51.80, SD=2.98), t=3.38, p=.00. They were also perceived by their parents to be at increased risk for attention problems and sluggish cognitive temperament. Lastly, children without sickle cell disease were perceived by their parents to be at increased risk for poor academic achievement.

Table 9

Primary Caregivers: Academic Achievement in Children with and without Sickle Cell Disease

	Children with SCD		**Children without SCD**		***t***	***p***
	M	***SD***	***M***	***SD***		
Thought Problems	52.29	4.24	51.33	2.93	1.46	.15
Attention Problems	55.6	5.82	52.09	3.40	3.40	.00*
ADHD	54.91	5.49	51.8	2.98	3.38	.00*
Academic Achievement	42.16	11.20	50.82	7.39	-4.55	.00*
Sluggish Cognitive Tempo	54.67	5.16	46.78	8.68	5.27	.00*

Note: *p<.05

CHAPTER 7

Discussion

Numerous studies have examined the psychosocial functioning and academic achievement in children and adolescents with a chronic illness (Kelch-Oliver et al., 2007). Although most children adjust well, studies suggest that they are at increased risk for psychosocial dysfunction (Kelch-Oliver et al., 2007). Sickle cell disease, as with other chronic illnesses, has the potential to severely disrupt developmental tasks of adolescence. Unpredictable and recurrent pain, delayed growth and sexual maturation, priapism, jaundice, and chronic fatigue are sensitive issues for adolescents with sickle cell disease (Kelch-Oliver et al., 2007). The most frequent psychosocial issues cited in the literature include poor self-concept, increased anxiety, depression, social withdrawal, aggression, and maladaptive peer relationships (Lutz et al., 2004).

Within the past two decades, there has been a growing interest in the psychosocial functioning and academic achievement in adolescents with sickle cell disease. Most of these studies have produced inconclusive results about the psychosocial functioning of children and adolescents with sickle cell disease (Gold et al., 2008). While some researchers have suggested that children and adolescents with sickle cell disease are at increased risk for psychosocial dysfunction when compared to healthy peers, other researchers have suggested that children and adolescents are not significantly different from their healthy peers (Gold et al., 2008; McClellan, Schatz, Sanchez, & Roberts, 2008). However, most of these studies had methodological limitations. For example, many of these studies were based entirely on structured interviews, clinical observations,

and psychiatric assessments. In addition, many of these studies did not include standardized measures of the behaviors and no comparison groups with test norms or controls. And lastly, many of these studies were conducted more than 20-years ago and are outdated.

The aim of the present study was to compare the psychosocial functioning and academic achievement in siblings with and without sickle cell disease. This researcher physically controlled for age, socioeconomic status, family environment, and parent education between siblings with and without sickle cell disease by using a case control design of siblings with and without sickle cell disease. This researcher also used well-validated measures to assess psychosocial functioning and academic achievement in this sample of adolescents with and without sickle cell disease. In addition, this researcher collected data not only from both siblings with and without sickle cell disease but also from their primary caregiver.

The results of this study support the hypothesis that psychosocial functioning and academic achievement is lower in siblings with sickle cell disease than in siblings without sickle cell disease. Overall, siblings with sickle cell disease reported more internalizing symptoms, somatic complaints and poorer social competence than their healthy siblings. They were perceived by their parents to be at increased risk for internalizing behaviors, poorer social competence and lower academic achievement. These results provide additional evidence that adolescents with sickle cell disease are at risk for poor psychosocial functioning and academic achievement. However, they also provide evidence that adolescents with sickle cell disease can function as well as

their healthy siblings with adequate family support, as all subjects had involved families and all children with sickle cell disease were involved in treatment.

This study improved on previous studies by using a case control design that has higher internal validity. This study also focused specifically on adolescents rather than children and adolescents. In addition, this study used convenience sampling and controlled dyads for age, socioeconomic status, family environment, and parent education. This researcher used an appropriate comparison group and measures that demonstrated adequate psychometric properties in African American adolescents. Lastly, this researcher was able to recruit a relatively robust sample size from the Sickle Cell Program at Children's National Medical Center in Washington, DC. This study built on prior research conducted at Children's Hospital of Philadelphia, St Christopher's Children's Hospital and Duke University Medical Center by clearly demonstrating that adolescents with sickle cell disease are at risk for poor psychosocial functioning and academic problems. These results have significant implications for social work practice.

Psychological Functioning

Recent studies examining the psychological functioning of adolescents with sickle cell disease have shown that they are at risk for psychological dysfunction (Ziadni, Patterson, Pulgaron, Robinson & Barakat, 2011; Simon et al., 2009; Trzepacz et al., 2004; Barakat et al, 2007; Kelch-Oliver et al., 2007). In this study, siblings with sickle cell disease reported clinically significant results on measures of internalizing behaviors, including withdrawal/depression, posttraumatic stress disorder, somatic complaints, and somatic problems. They reported more recurrent, unexplained pain,

headaches, and stomach aches and higher levels of sleep disturbance than their healthy siblings. They also report clinically significant results for anxiety and other related disorders, which supports the literature that adolescents with sickle cell disease are at increased risk for anxiety (Simon et al., 2009; Barakat et al., 2006; Lee et al., 1997; Lemanek et al., 1986).

Children with sickle cell disease were perceived by their parents to be at increased risk for internalizing behaviors, including withdrawal/ depression, affective problems, somatic complaints and somatic problems. However, they were not perceived by their parents to be at increased risk for anxiety. Although, some studies have shown high rates of depression and anxiety in children and adolescents with sickle cell disease, other studies have failed to show clinically significant levels of anxiety in these patients (Alao & Cooley, 2001). As a result, clinical evidence of anxiety and depression in adolescents with sickle cell disease still remains unclear (Pao & Bosik, 2011; Simon et al., 2009).

While some studies have reported that adolescents with sickle cell disease are at risk for psychological dysfunction, particularly externalizing behaviors, other studies have shown that they are not at greater risk for externalizing behaviors when compared with normative samples and healthy controls (Simon et al., 2009; Gold et al., 2008, Trzepacz et al., 2004; Thompson et al., 1998). In this study, siblings with sickle cell disease did not differ significantly from their healthy siblings on measures of externalizing behaviors. Their parents reported similar results. These findings suggest that vaso-occlusive crisis, chronic fatigue and smaller physical stature may lead to more passive behavior in these adolescents (Noll et al., 2007; Trzepacz et al., 2004).

Over the past several years, researchers have begun to rely on conceptually based theoretical models to explain psychological functioning. Wallander and Varni (1992) disability-stress-coping model uses Lazarus and Folkman's (1984) cognitive appraisal concepts to explain psychological adjustment in children with chronic illnesses (Manuel, 2004). According to Lazarus and Folkman (1984), person and situation factors may influence the appraisal process by determining if a particular situation is stressful or threatening. For adolescents with sickle cell disease, primary appraisal is the evaluation of whether sickle cell disease presents stressors or demands that threaten their well-being (Lazarus & Folkman, 1984). Secondary appraisal is the evaluation of whether they have the resources to cope with their illness (Lazarus & Folkman, 1984). Some studies have found primary appraisal to be linked to positive adjustment in adolescents with sickle cell disease (Casey et al., 2000; Thompson et al., 1998; Brown et al., 1993).

According to Wallander and Varni (1992), sickle cell disease is viewed as a potential threat to adolescents' psychological adjustment (Casey et al., 2000). Protective or resistance factors are associated with better psychological functioning (Simon et al., 2009; Palermo et al., 2008; Kelch-Oliver et al., 2007; Burlew et al., 2000). They include such protective factors as (a) personal characteristics (e.g., age, gender, temperament), (b) family factors (e.g., social support, socioeconomic status, adjustment of family member), and (c) stress processing (e.g., cognitive appraisal coping). Studies have found that children from low socioeconomic status families experience more adjustment problems than children from high socioeconomic status families (Panepinto et al., 2009; Kelch-Oliver et al., 2007). Specifically, they are higher risk for externalizing behaviors than

children for high socioeconomic status families. In this sample, family incomes and parent education were higher than most normative samples, which might explain our results on measures of externalizing behaviors (Burlew et al., 2000). As a result, protective factors such as sociodemographic characteristics may play a pivotal role in fostering positive psychological adjustment in these adolescents.

Over the last decade, researchers have begun to study the role of family functioning on psychosocial adjustment in children and adolescents with sickle cell disease (Alderfer & Hodges, 2010, Barakat, Lutz, Nicolaou, & Lash, 2005). Adaptive family relationships have been linked to positive psychosocial adjustment in these children and adolescents (Herzer, Godiwala, Hommel, Driscoll, Mitchell, Crosby, Piazza-Waggoner, Zeller, & Modi, 2010). Prior research has shown that children and adolescents from cohesive families report better psychosocial functioning than children and adolescents from less cohesive families (Pinckney & Stuart, 2004). In addition, family competence and adaptability have been linked to improved emotional and behavior functioning in these patients (Pinkney & Stuart, 2004). And lastly, higher levels of family conflict and lower levels of family support have been linked to more symptoms of depression and anxiety and lower social competence (Pinckney & Stuart, 2004).

A growing body of evidence suggests that sibling relationships may also have a profound impact on the psychosocial adjustment of children and adolescents with sickle cell disease. Emotional intimacy may serve as a protective factor in sibling relationships (Soli, McHale, & Feinberg, 2009). According to the literature, positive sibling relationships may mediate illness-related effects and serve as a protective factor in the

social and emotional development of adolescents with sickle cell disease (Drotar & Crawford, 1985). Consequently, the relationship may instill in the adolescents with sickle cell disease a sense of self-worth and competence. In addition, researchers have found that sibling relationships may foster positive self-esteem and contribute to self-efficacy (Soli et al., 2009). However, hostile and aggressive sibling relationships may increase adolescents' risk for internalizing and externalizing behaviors and lower social competence (Soli et al., 2009). As a result, sibling relationships may increase adolescents risk for peer rejection and contribute to feels of loneliness and isolation.

The data support the hypothesis that siblings with sickle cell disease are at higher risk for poor psychosocial functioning and academic achievement than their healthy siblings. They reported more symptoms of anxiety, depression, withdrawal and somatic complaints. They also reported more social problems and lower social competence. However, the mean levels did not fall outside the normal range, suggesting that adolescents in this sample of patients with sickle cell disease cope relatively well with their illness. They reported similar results for academic achievement. These results highlight the importance of family functioning on the psychosocial adjustment in adolescents with sickle cell disease. Although, this study did not specifically examine family functioning, it might explain why siblings with and without sickle cell disease did not report results outside the normal range. As a result, the data from this study indicate that it is possible for adolescents with sickle cell disease to cope as well as their healthy siblings with adequate family support.

The results from this study have significant implications for social work practice

in adolescents with sickle cell disease. According to the literature, social workers should routinely screen for psychological adjustment in this patient population. The Beck Depression Inventory, Rosenberg Self-Esteem Scale, the Child Behavior Checklist, the Depression Self-Rating Inventory, the Rathus Assertiveness Schedule, and the Youth Self-Report have all been shown to be effective tools for assessing psychosocial functioning (Pinckney & Stuart, 2004). Adolescents who are at increased risk for poor psychological functioning should be referred for psychological support services. Psychoeducation is one treatment modality that has been shown to be effective in improving psychological functioning in adolescents with sickle cell disease. The focus of treatment should be on enhancing adolescents' self-esteem and self-efficacy skills through social support and assertion skills training (Kelch-Oliver et al., 2007; Pinckney & Stuart, 2004). Research has shown a direct correlation between social support and better coping in adolescents with a sickle cell disease (Anie, 2005). Both individual and group therapy can also be effective in improving psychological functioning.

A growing body of literature has shown that cognitive behavioral therapy and acceptance and commitment therapy are effective treatment modalities in improving psychological functioning in adolescents with sickle cell disease (Pao & Bosik, 2011; Masuad et al., 2011; Kelch-Oliver et al., 2007). Cognitive behavioral therapy and acceptance and commitment therapy work by modifying maladaptive or dysfunctional behaviors (Keefe, Dunsmore, & Burnett, 1992). These therapies focus on adolescents' cognitions (thoughts, beliefs, attitudes), emotions (fear of their disease), and behaviors (activity avoidance due to their disease) and their impact on psychological functioning

(Keefe et al., 1992). These therapies teach adolescents about their illness and its relation to cognitive, affective, and behavioral dysfunction, and how to think differently about their illness. Emphasis is placed on the role adolescents have on managing their condition (Beissner et al., 2009). As a result, they learn new skills for coping effectively with their illness (Keefe et al., 1992).

Social Competence

Interpersonal relationship is an important factor in the psychosocial functioning of children and adolescents with chronic illness. Peer relationships are an important source of emotional support in these children and adolescents. Wallander and Varni (1992) suggested that peer relationships are associated with better social adjustment. Studies have shown that children and adolescents with a chronic illness may be at significant risk for poor social competence.

Over the last 20-years, researchers have begun to examine social competence and interpersonal functioning in adolescents with sickle cell disease (Ziadni et al., 2011). Many of these studies have shown that adolescents with sickle cell disease are at increased risk for social difficulties (Ziadni et al., 2011). Compared with healthy peers, these adolescents may experience feelings of loneliness and isolation. They may be more vulnerability and less self-accepting of their medical condition. They may also be socially inhibited. As a result, they may have fewer friends and social contacts outside of school (Anie, 2005). However, many of these studies had methodological limitations. They focused on the perceptions of parents, usually mothers, and not the adolescents (Rodrigue et al., 1996). They also lacked appropriate comparison groups and

standardized measurements (Rodrigue et al., 1996). Therefore, many of these studies produced inconclusive results.

In this study, siblings with sickle cell disease reported more social problems and lower social competence than their healthy siblings. They reported more social withdrawal and lower self-esteem than their healthy siblings. They were also perceived by their parents to be at increased risk for poor social competence. However, healthy siblings were also perceived by their parents to be at increased risk for poor social competence. Studies have shown that healthy siblings may experience behavioral problems, school problems, feelings of parental rejection and somatic symptoms (Vermaes, van Susante, & van Bakel, 2012). They may also experience feelings of confusion, guilt and lower self-concept. As a result, these findings are consistent with the current literature that suggests that adolescents with and without sickle cell disease, where one has sickle cell disease, are at higher risk for poor social competence compared with normative samples (Edwards et al., 2005; Rodrigue et al., 1996).

Parental distress may also be a predictor of social problems and low social competence in adolescents in sickle cell disease. Studies have shown that many parents of children with sickle cell disease view their child's illness as serious, its management as a burden, and experience a lack of control over the illness (Williams, 1997). Although most studies indicate that parental distress in parents of children with sickle cell disease decreases over time, some parents continue to experience high levels of distress throughout the child’s development. While this study did not assess parental distress, it might explain the high levels of poor social competence and interpersonal

functioning in this sample of adolescents with sickle cell disease. Parental distress has been associated with an overprotective style of parenting which may lead to higher internalizing and externalizing behaviors, lower social competence, low self-esteem, and withdrawal in adolescents with sickle cell disease and their siblings. As a result, adolescents may lack experiences necessary for healthy psychosocial development.

Physiologic complication may also have a significant impact on social competence in adolescents with sickle cell disease. Central nervous system complications, such as silent and overt strokes, may impair adolescents' social functioning. They may have difficulty developing age-appropriate peer-relationships and understanding social processes (Baker et al., 2012). Some studies have found these impairments increase adolescents' risks for social problems and lower social competence (Baker, Niec, & Meade, 2012). This sample of adolescents with sickle cell disease reported problems with interpersonal relationships. They reported poor peer relationships and less peer and teacher affiliation than their healthy siblings. They also reported aggressive and hostile peer relationships.

Vaso-occlusive crisis has also been shown to have an adverse effect on social competence and interrpersonal relationships in adolescents with sickle cell disease. It is the most common complication of sickle cell disease. The age of onset, frequency, and duration of vaso-occlusive crises vary according to the literature (Hocking & Lochman, 2005). The average adolescent with sickle cell disease experiences 1-2 vaso-occlusive crises a month, each lasting 4-6 days, and 1-2 hospital admissions and emergency room visits a year (Hocking & Lochman, 2005). It has been linked to a wide range of

adjustment problems, including, less social confidence and more affective and behavioral problems in adolescents with sickle cell disease (Hocking & Lochman, 2005). As a result, neurological complications and vaso-occlusive crisis might explain these results in this sample of adolescents with sickle cell disease.

These data have significant clinical implications for social work practice. Social workers should routinely screen for social competence. Studies have shown that social support enhancement is critical to optimize social competence in adolescents with sickle cell disease. Because adolescents spend most of their time with peers, peer-group support has been shown to be a particularly effective tool in improving social competence. Studies have shown that peer support groups offer a buffering affect and help build self-esteem, personal identify, and social and moral development. In addition, studies have found that social skills training for adolescents with sickle cell disease offer similar benefits (Noll et al., 2007; Barakat et al., 2003). Social skill training that includes multiple components such as problem solving, modeling, and social perception may be especially effective for improving social competence in adolescents with sickle cell disease (Barakat et al., 2003). However, more longitudinal research is needed to assess the efficiency of these interventions over time.

Academic Achievement

The number of studies examining academic achievement in children and adolescents with sickle cell disease has dramatically increased over the past decade. In most of these studies, psychometric tests and academic skills tests have been used to assess academic achievement in adolescents with sickle cell disease (Noll et al., 2001; Wang et al., 2001). As a result, there is increasing evidence to suggest that adolescents with sickle cell disease are at significant risk for cognitive impairment. Neurologic complications, including silent and overt stroke, can result in deficits in cognitive and social functioning (McClellan et al., 2008; Noll et al., 2001). Studies suggest that older children with sickle cell disease show more cognitive deficits than younger children with sickle cell disease (Anie, 2005; Schatz, Finke, Kellett & Kramer, 2002). There is also some evidence to suggest that children with a history of overt strokes may perform worse academically than children who have had silent infarcts (Anie, 2005; Schatz et al., 2002). In addition, children who have had neurologic complications may have learning deficits in reading and mathematics (Schatz et al., 2002). Although, most studies examining academic achievement in adolescents with sickle cell disease demographically matched for age, ethnicity, and race, the best comparison groups are siblings. They have greater similarity (Schatz et al., 2002). In addition, many of these studies used small sample sizes (Noll et al., 2001).

In this study, siblings with sickle cell disease reported more thought problems than their healthy siblings. They also reported more attention and memory problems, lower information processing abilities, and more language and visual-spatial deficits than

their healthy siblings. They also reported deficits in executive functioning. In addition, they were perceived by their parents to be at increased risk for attention problems, sluggish cognitive temperament and attention deficit hyperactivity disorder than their healthy siblings. Although central nervous system impairments were not assessed as part of this study, they might explained the high rate of poor academic achievement in this sample of sibling with sickle cell disease.

In addition to central nervous system impairments, poor academic achievement might also be explained by vaso-occlusive crisis and chronic anemia. Studies have shown that 65% of children and adolescents with sickle cell disease experience five to seven vaso-occlusive crises per year, lasting more than 2-weeks (Hoff, Palermo, Schluchter, Zebracki, & Drotar, 2006). These children and adolescents may miss an average of 20-30 days from school each year (Peterson et al., 2012; Schwartz, Radcliffe, & Barakat, 2009). Chronic anemia has also been associated with deficits in cognitive functioning, verbal ability and working memory (Bernaudin et al., 2000). As a result, complications of sickle cell disease may increase adolescents risk for poor academic achievement (Day & Chismark, 2006).

These results have significant implications for social worker practice. The literature suggests that social workers should routinely screen for academic risk factors, including attention and concentration problems, in adolescents with sickle cell disease. This could be done during annual comprehensive visits using measures such as the Continuous Performance Test or Trails A and B (Noll et al., 2001). Close monitoring will help identify those at risk so that interventions can be developed to reduce their risk

for poor academic achievement. Social workers should also work closely with school nurses to bridge the gap between healthcare providers and teachers (Day & Chismark, 2006). Nurses should be encouraged to organize workshops and trainings about sickle cell disease for teachers. These workshops and trainings should include information about sickle cell disease, common complications associated with sickle cell disease, and information about special education services under the Individuals with Disabilities Education Act (Day & Chismark, 2006). Children and adolescents with sickle cell disease are eligible for classroom accommodations, transportation, school health services, speech and language services, physical and occupational therapy, and psychological services (Day & Chismark, 2006). They are also eligible for additional time to complete assignments and tests, tutoring, and homebound instruction (Day & Chismark, 2006). Furthermore, they are eligible for neuropsychological or cognitive educational testing. And lastly, teachers should be cognizant of poor psychological and social functioning in adolescents with sickle cell disease. They should be sensitive to issues of peer acceptance and self-esteem and assist with reintegration after absences (Bonner et al., 1999). School conferences should focus on fostering communication among healthcare providers, teachers, patients and parents (Bonner et al., 1999).

Strengths and Limitations of the Study

This study has numerous strengths as well as limitations. This is a well-designed study that contributes to the current literature about psychosocial functioning and academic achievement in adolescents with sickle cell disease. The study uses a case control design to assess psychosocial functioning and academic achievement in

adolescents with sickle cell disease. Case control studies have higher internal validity. The study also uses a demographically matched comparison group and it controls for ages, parent education, and family environment in the dyad. It also uses parental report of psychosocial functioning and academic achievement. Furthermore, it uses the Youth Self-Report and the Child Behavior Checklist, which are both well validated measures to assess psychosocial functioning and academic achievement in adolescents with sickle cell disease. Lastly, 133 adolescents with and without sickle cell disease and their caregivers completed the study. Compared to prior studies, this is a robust sample size that further documents that adolescents with sickle cell disease are at increased risk for poor psychosocial functioning and academic achievement. This study reinforces the importance of appropriate comparison groups that controls for age, socioeconomic status, family environment, and parent education.

Although the sample size is large in the context of other studies of those with sickle cell disease, it is still a relatively small sample. The study was conducted at one site. This researcher used convenience sampling of those in treatment to recruit siblings with and without sickle cell disease and their primary caregivers which may have biased our sample toward higher functioning adolescents with sickle cell disease. Adolescents who attend clinic regularly often cope better with their illness and are more likely to seek medical attention. In addition, their parents tend to be more educated and knowledgeable about sickle cell disease and their child's illness. As a result, they are more likely to experience better psychosocial functioning.

The Youth Self Report and Child Behavior Checklist have been widely used by

researchers in clinical and research settings to examine the psychosocial functioning in children and adolescents with a chronic illness, but it is important to recognize their limitations. The Youth Self Report and Child Behavior Checklist are well validated measures, but they were not specifically designed to assess psychosocial functioning and academic achievement in adolescents with sickle cell disease (Lambert, Rowan, Lyubansky, & Russ, 2002). As a result, the cultural and content validity must be taken into consideration for possible bias and conclusions (Lambert et al., 2002). For example, somatic symptoms could inflate psychopathology in adolescents with sickle cell disease. In addition, these measures have the potential to overlook subtle adjustment problems that are often experienced by adolescents with chronic medical conditions. In addition, although this researcher used the best measures available to assess psychosocial functioning and academic achievement in this sample of adolescents with sickle cell disease, future studies would be better served by employing specifically measures designed to assess psychosocial functioning and academic achievement in adolescents with sickle cell disease. Finally, while this study has significantly improved upon previous research that has examined the psychosocial functioning and academic achievement in adolescents with sickle cell disease by using a multi-informant methodology, future studies may benefit from the inclusion of behavioral observations and teacher reports.

Lastly, these findings are cross-sectional. While the study provides evidence that adolescents with sickle cell disease are at increased risk for poor psychosocial functioning and academic achievement, longitudinal studies are necessary to understand

the impact of sickle cell disease across the lifespan. Most adolescents with sickle cell disease now survive well into adulthood. As a result, it is important for researchers to engage in well-designed longitudinal studies with appropriate comparison groups to understand the impact of sickle cell disease from adolescence to adulthood.

Despite these limitations, the study makes a significant contribution to the growing body of literature about the psychosocial functioning and academic achievement in adolescents with sickle cell disease. Results of this study provide strong evidence that adolescents with sickle cell disease are at risk for poor psychosocial functioning and academic achievement, but it must be recognized that these analyses were all at the bivariate level. Further analyses at the multivariate level are needed. These findings also provide strong evidence that it is possible for siblings with sickle cell disease to cope as well as their healthy siblings with adequate family support. More well-designed studies are needed to examine the psychosocial functioning and academic achievement in this population. Lastly, this study highlights an urgent need for researchers to develop, evaluate and implement effective interventions to prevent poor psychosocial functioning and academic achievement in adolescents with sickle cell disease.

Conclusion

Sickle cell disease is a chronic and debilitating illness that can have a profound impact on adolescents' psychosocial functioning. Although many adolescents with sickle cell disease adjust well, other adolescents are at increased risk for adjustment problems (Hocking & Lochman, 2005). Complications of sickle cell disease such as unpredictable pain, pneumococcal infections, anemia, delayed growth and puberty, splenic

sequestration, stroke, leg ulcers and bone lesions increase adolescents' risk for poor psychosocial adjustment and academic achievement. Within the last decade or so, more researchers have begun to examine the psychosocial functioning and academic achievement in adolescents with sickle cell disease. However, many of these studies have methodological limitations and have produced inconclusive results. This well designed study provides additional evidence that adolescents are at increased risk for poor psychosocial functioning and academic achievement. These results are consistent with a rapidly growing body of literature indicating that these adolescents are at significant risk for poor psychosocial functioning and academic achievement. However, the results also provide strong evidence that it is possible for siblings with sickle cell disease to cope as well as their healthy siblings with family support. Social workers should screen for poor psychosocial functioning during clinic visits. Interventions should be offered as standard care for all adolescents with sickle cell disease. They should focus on identifying and treating maladaptive behaviors. Cognitive behavior therapy and psychoeducation have shown to be effective treatment modalities for improving psychosocial functioning in adolescents with sickle cell disease. Lastly, given the importance of psychosocial functioning and academic achievement in adolescents with sickle cell disease, more longitudinal studies are needed to understand psychosocial functioning in adolescents with sickle cell disease across the lifespan.

APPENDIX

Lisa Thaniel, MSW, LICSW
Doctoral Social Work Student
School of Social Policy and Practice
University of Pennsylvania
3701 Locust Walk
Philadelphia, PA 19104-6214

Date:

Dear_____,

My name is Lisa Thaniel. I am a doctoral student in social work at the School of Social Policy and Practice at the University of Pennsylvania. I would like to invite you to participate in a research study at Children's National Medical Center.

I am conducting a research study comparing the psychosocial functioning and academic achievement in siblings with and without sickle cell disease. I am recruiting siblings with and without sickle cell disease who are between the ages of 12 and 18 and their primary caregivers to be in the research study. If you agree to participate in the study, you will be required to complete the Children's National Medical Center consent or assent form and a series of surveys on Survey Monkey. The surveys should not take more than 2 hours to complete. Your participation in the study is completely voluntary; you are free to withdraw your participation from the study at any time. There is a small chance that some of the questions may make you feel uncomfortable. You do not have to answer those questions if you do not want to. In fact you do not have to answer any questions that you choose not to answer. For participating in the study, your family will be compensated with a $25 gift card, a $5.00 meal ticket to the cafeteria, and parking validation (if needed).

I hope you will consider being in this research study. If you are interested in this study, please contact me at (202) 476-3555 or lthaniel@childrensnational.org.

Thank you for your time.

Sincerely,

Lisa Thaniel, MSW, LICSW
Doctoral Social Work Student

Telephone Script

Hello, my name is Lisa Thaniel. I am a doctoral student in social work at the School of Social Policy and Practice at the University of Pennsylvania. May I please speak to_____?

If the sibling without sickle cell disease is there- continue with the script.
If the sibling without sickle cell disease is not there-ask when would be a good time to speak with _____?

I am conducting a study comparing the psychosocial functioning and academic achievement in siblings with and without sickle cell disease. I am recruiting siblings with and without sickle cell disease who are between the ages of 12 and 18 and their primary caregivers to be in the study. I recently spoke to your mother/father/caregiver and sibling with sickle cell disease in clinic. Your mother/father/caregiver gave me permission to contact you so that I might ask you to participate in the study. Is this a good time to talk?

If this is a good time to talk-continue with the script.
If this is not a good time to talk-ask when would be a good time to call again.

If you agree to participate in the study, you will be asked to complete the Children's National Medical Center consent or assent form and a series of surveys on Survey Monkey. The surveys should not take more than 2 hours to complete. Your participation in the study is completely voluntary; you are free to withdraw your participation from the study at any time. It is important to note, there is a small chance that some of the questions may make you feel uncomfortable. You don't have to answer those questions if you don't want to. In fact you don't have to answer any questions that you choose not to answer. And that is fine. Your information will be kept confidential and will only be seen by researchers at the University of Pennsylvania and Children's National Medical Center. For participating in the study, your family will be compensated with a $25 gift card, a $5.00 meal ticket to the cafeteria, and parking validation (if needed).

Do you have any questions?

Would you be willing to participate?

If no: Thank you for your time and stop the recruitment process.
If yes: Great! Let me give you some important information about the study. Please feel free to stop me any time if you have any questions.

The title of the study is: A Case Control Study: The Psychosocial Functioning and Academic Achievement in Adolescents with and without Sickle Cell Disease. To access

the surveys, you will need to log into Survey Monkey. Again the address is: www.surveymonkey.com. The login word is _____ and the password is _____. If you are between the ages of 12 and 17, you will be asked to electronically sign the Children's National Medical Center assent form and if you are 18 years old, you will be asked to electronically sign the Children's National Medical Center consent form. Survey Monkey will then direct you to the surveys.

Do you have any questions?

No follow-up is required and the gift card will be sent to you and your family upon completion of the study. In addition, I will contact you to conduct a debriefing if you exit the study early or upon completion of the study (within 24 hours) to see if you have any questions or concerns.

Do you have any questions?

Again, my name is Lisa Thaniel, LICSW. I am a doctoral student in social work at the School of Social Policy and Practice at the University of Pennsylvania and I am the co-investigator for the study. Emily Meier, MD is the Principal Investigator. If you have any questions or concerns, please feel free to contact me at lthaniel@childrensnational.org or (202) 476-3555.

Thank you for your consideration of involvement in this study.

Goodbye

Debriefing Script

Hello, my name is Lisa Thaniel. I am a doctoral student in social work at the School of Social Policy and Practice at the University of Pennsylvania. May I please speak to_____?

If the siblings with and without sickle cell disease and their caregivers are there- continue with the script.
If the sibling with and without sickle cell disease and their caregivers are not there-ask when would be a good time to speak with _____? I will conduct a debriefing with the siblings and caregivers individually in person or via telephone.

Thank you for taking time to participate in our study. The purpose of the study is to compare the psychosocial functioning and academic achievement in adolescents with and without sickle cell disease. A substantial body of literature provides strong evidence that chronically ill adolescents are at risk for psychosocial dysfunction. However, few current studies have clearly demonstrated that adolescents with sickle cell disease are at risk for psychosocial dysfunction and problems with academic achievement.

Do you have any questions or concerns about the study?

If participation in our study has caused you to feel uncomfortable in any way, I encourage you to contact me directly at (202) 476-3555. If you need immediate psychological services, I encourage you to take advantage of the confidential counseling services offered by Dr. Amanda Thompson, clinical psychologist in the Hematology-Oncology Department. I also encourage you to use the psychiatric liaison on-call or the emergency department at Children's National Medical Center. (If the caregiver feels uncomfortable in any way, I will refer the caregiver to the Washington Hospital Center for psychological services). In addition, if you have a concern or complaint about the study, I encourage you to call the Principal Investigator at (202) 476-2800 or the Chief Academic Officer of the Children's National Medical Center at (202) 476-5000.

Thank you again.

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