THE EFFECTS OF FAMILY FUNCTIONING ON ACADEMIC ACHIEVEMENT IN CHILDREN WITH SICKLE CELL DISEASE

by

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Sickle cell disease (SCD) is a complex group of genetic blood disorders that currently affects 90,000-100,000 Americans primarily of African descent. SCD leads to physiological and psychosocial distress. In relation to school, youth with SCD are at high risk of poor academic outcomes, including: low scores on tests of academic achievement, and increased risk of poor grades, special education, and grade retention. There is a paucity of literature on family functioning's effect on academic functioning in youth with SCD. Poor family functioning in youth with SCD has been related to many other psychosocial outcomes, such as behavior problems, poor mental health and quality of life. The current study examined whether family functioning is directly related to the academic outcomes of youth with SCD, and investigated whether family functioning moderates the relationships between disease severity, SES, age, and academic outcomes, using simultaneous multiple regression models. The current study utilized data collected at the beginning of phase three of the Cooperative Study of Sickle Cell Disease (CSSCD), consisting of 198 youth aged 6 to 16 years. Family functioning was evaluated using the Family Environment Scale, academic achievement by broad reading and math scores from the Woodcock-Johnson Revised Tests of Academic Achievement, and school competence by the School Competence Scale of the CBCL. Results indicate that family functioning variables were

neither directly related to academic outcomes, nor did they moderate the relationship between academic outcomes and other factors. Results also indicated that IQ as measured by the FSIQ of the WISC-III was the most powerful predictor of academic functioning. Limitations and clinical implications are discussed.

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TABLE OF CONTENTS

LIST OF TABLES

CHAPTER I: INTRODUCTION

Youth with sickle cell disease (SCD) are at high risk of poor academic outcomes. In comparison to healthy children, they are more likely to score low on tests of academic achievement, to earn poor grades, to receive special education services, and to be retained a grade (Boulet, Yanni, Creary, & Olney, 2010; Peterson, Palermo, Swift, Beebe, & Drotar, 2005). Factors that have been linked to poor academic outcomes in children with SCD include poor neurocognitive functioning primarily due to stroke, high disease severity, and low socioeconomic status (SES). However, one factor that has been largely ignored in relation to the academic functioning of children and adolescents with SCD is family functioning. Poor family functioning in youth with SCD has been related to behavior problems (Thompson et al., 2003), poor mental health (Kell, Kliewer, Erickson, & Ohene-Frempong, 1998) and quality of life (Barakat, Lutz, Nicolaou, & Lash, 2005). Evidence also links lower family functioning in healthy youth to worse academic performance (King, 1998), and preliminary research has linked aspects of family functioning to academic achievement in youth with SCD (Barbarin, Whitten, Bond, & Conner-Warren, 1999).

The purpose of the current study is to examine how functioning in families with children with SCD impacts the academic outcomes of these children. The following review of literature provides an overview of pediatric SCD, describes the academic outcomes of children with SCD, describes factors known to influence these academic outcomes, and reviews family functioning as a possible influence on the academic outcomes of children with SCD.

Sickle Cell Disease

Sickle cell disease (SCD) is a broad term for a group of chronic, genetic blood disorders affecting the chemical structure of red blood cells. The disease currently affects 90,000 to

100,000 Americans (National Heart Lung and Blood Institutes, 2012). In the United States, it is primarily seen in people of African descent, but is also seen in people of Latino and Mediterranean descent. SCD occurs in 1 in 500 African-American births and 1 in 36,000 Latino births. The genetic trait for passing SCD on to future offspring, known as sickle cell trait, occurs in 1 in 12 African-Americans (Centers for Disease Control and Prevetion [CDC], 2012).

For people with SCD, high concentrations of sickled hemoglobin (HbS) causes deoxygenated red blood cells to polymerize and form stiff sickled, or "C", shapes rather than remaining round and flexible, as do typical red blood cells (Steinberg, 2005). These malformed blood cells cause vasoocclusions by adhering to each other and to the walls of blood vessels. These cells are also less efficient in carrying oxygen and die more quickly than typical red blood cells. The poor functioning of the sickled red blood cells can cause a range of complications, including acute and chronic pain, anemia, severe infections, acute chest syndrome, stroke, organ failure, vision loss, and leg ulcers.

Sickle cell anemia (HbSS), the most common form of SCD, is caused by having two genes for HbS, one from each parent (Bonner, Hardy, Ezell, & Ware, 2004). Other forms of SCD occur when a person inherits an HbS gene and another abnormal hemoglobin gene. For example, the two most common forms of SCD behind HbSS are sickle cell-hemoglobin C disease (HbSC) and sickle cell beta-thalassemia (HbSβ), which comes in two forms: HbSβ° or HbSβ$^+$ (Redding-Lallinger & Knoll, 2006). Notably, disease severity can roughly be predicted through SCD genotype, as HbSS and HbSβ° are considered more severe forms of SCD than HbSC and HbSβ$^+$ because they are associated with more SCD-related complications (Sebastiani et al., 2007).

Since the discovery of SCD in 1949, medical advances have dramatically improved the prognosis of people with SCD (Bunn, 1997). SCD was once considered a pediatric disease, with

a life expectancy of age 14 years in 1973 (Platt et al., 1994). However, people with SCD now live into adolescence and adulthood, with a life expectancy of 42 and 48 years for men and women with HbSS respectively, and 60 and 68 years for men and women with HbSC respectively. The increased longevity of people with SCD makes a focus on their academic outcomes more important because academic outcomes are directly related to adult economic prospects (Cheeseman Day & Newburger, 2002).

Academic Outcomes of Children with SCD

The majority of studies looking at academic outcomes in children with SCD provide evidence that youth with SCD have worse academic outcomes than their healthy peers. Boulet, Yanni, Creary, and Olney (2010) studied black children aged 0–17 years, 192 with SCD and 19,335 without SCD. The researchers found that children with SCD were more likely to receive special education services than black children without SCD. In a study of 72 children with SCD aged 5-17 years, Peterson, Palermo, Swift, Beebe and Drotar (2005) found that children with SCD performed below average on achievement and IQ tests. The researchers also found that 42% children with SCD reported having disease-related difficulties participating in school, 36% had been retained at least one grade, 28% had individualized education plans, and 35% had missed 20 or more days of school. Fowler et al. (1988) studied 28 children with HbSS aged 6 -17 years compared to 28 healthy children matched on race, age, sex, and SES. They found that children with HbSS performed worse on measures of reading and math achievement than controls. This finding is consistent with results from Swift and colleagues' (1989) study of 28 children with HbSS aged 7 – 16 years and 21 of their healthy siblings, which found that children with HbSS underperformed in comparison to the healthy controls on tests of memory, reading achievement and math achievement. Lastly, Brown, Buchanan, Doepke, and Eckman's (1993)

study of 70 children with SCD aged 3 – 17 years and 18 healthy siblings found that children with SCD had lower overall academic achievement scores on the K-ABC Achievement Battery and the Basic Achievement Skills Individual Screener (BASIS) than their healthy siblings. The following sections review the literature on factors believed to contribute to poor academic outcomes in children with SCD.

Factors Influencing Academic Outcomes in Children with SCD

Neurological Functioning. The primary factor believed to account for the problems seen in academic outcomes in children with SCD is poor neurocognitive functioning. Studies have found that children with SCD score significantly lower on tests of neurocognitive functioning compared to healthy controls (Brown et al., 1993; Fowler et al., 1988; Wasserman, Williams, Fairclough, Mulhern, & Wang, 1991) and that children with SCD have lower IQ scores than their healthy siblings (Swift et al., 1989; Wasserman et al., 1991).

Researchers hypothesize that the difficulties children with SCD evidence in neurocognitive functioning are primarily a result of cerebrovascular accidents or strokes. Stroke is a clinical syndrome that results in an insufficient supply of blood to a part of the brain (Markus, 2003). A stroke that occurs with clinical symptoms, such as paralysis, headache, confusion, loss of vision, and loss of motor control, is referred to as an overt stroke. In contrast, a "silent" stroke is defined as the presence of a structural defect in the brain, as seen through modern imaging techniques, such as MRI, with the absence of clinical symptoms of stroke (Pegelow et al., 2001). Children with SCD are 333 times more likely to have a stroke than healthy children; with overt stokes occurring in 11% of youth with HbSS before age 20, and silent strokes occurring in 10-30% of people with SCD (Verduzco & Nathan, 2009).

Both overt and silent strokes have been linked to poor neurocognitive and academic functioning in children with SCD. Schatz, Brown, Pascual, Hsu, and DeBaun's (2001) study of 19 children with SCD who had experienced silent stroke, 45 children with SCD but no stoke, and 18 of their healthy siblings aged 8-15 years found that children with SCD and evidence of stroke had lower IQ scores and lower academic achievement scores than those children with SCD and without evidence of stroke. Wang et al. (2001) conducted a larger study using 373 children with SCD aged 6-18 years from the Cooperative Study of Sickle Cell Disease (CSSCD) and found that children with HbSS and silent strokes had significantly lower achievement scores in both math and reading, and lower verbal and performance IQs than children with HbSS and normal MRIs. In addition, studies have found that youth who have experienced overt strokes tend to have lower IQ scores and lower academic achievement scores than children who have had silent strokes and children that have no history of stroke (Brown et al., 2000; Daly, Kral, & Brown, 2008; Kral, Brown, & Hynd, 2001). Of note, academic difficulties have also been noted in children with SCD who have no evidence of stroke. For example, Schatz et al. (2001) studied 19 children with SCD and silent infarct, 45 children with SCD and no history of infarct, and 18 healthy siblings and found that 27% of children with SCD and no evidence of stroke had either been retained or required remedial school services whereas only 6% percent of their healthy siblings had poor academic achievement. Youth with SCD have a high risk of having their neurocognitive functioning compromised by overt or silent stroke, which contributes to low IQ and achievement scores among these children. Poor neurocognitive functioning, however, does not explain poor academic achievement in all youth with SCD, and additional contributions to poor achievement have been explored.

Disease Severity. Another factor hypothesized to impact academic functioning in children with SCD is disease severity. Disease severity has been measured in several ways, including: sickle cell genotype, number of pain episodes, hospitalizations, hemoglobin levels, and complications related to SCD. In a study using a sample of 1,772 children with SCD aged 5-15 years, more pain episodes were associated with poor school functioning (Dampier et al., 2010). Eaton, Haye, Armstrong, Pegelow, and Thomas (1995) studied 21 children with HbSS and no history of CVA and compared them on frequency of hospital stays. They found that children with a high frequency of hospitalization for pain missed significantly more days of school than children with a low frequency of hospitalization. Notably, both groups (i.e., those with a high and low frequency of hospitalizations) had less than a C average in school. Mayes, Wolfe-Christensen, Mullins, and Cain (2011) found that greater disease severity, as measured by a composite of the number of emergency room visits, hospitalizations, days hospitalized, and average hemoglobin levels, was associated with special education placement and higher parental concern for school performance. Nettles (1994) compared the norm referenced academic achievement test scores of 17 children with HbSS, 15 children with HbSC, and 34 healthy children aged 6-16 years. Findings indicated that the groups with SCD had lower achievement scores than the healthy groups; however there was no significant difference between the HbSS group and the HbSC group. In fact, although the HbSS group trended towards having worse attendance than the HbSC group, the HbSC group trended towards having worse reading scores than the more severe HbSS group. Nettles' findings may differ from the findings of other research because she only used SCD genotype to measure disease severity, whereas other studies used composites of symptomatology and/or healthcare utilization to measure severity. Taken

together, the majority of research indicates that children with more SCD related complications perform worse academically than children with less SCD related complications.

Age and SES. Additional factors believed to impact academic achievement in children with SCD are age and socioeconomic status (SES). Evidence indicates that as youth with SCD grow older, they fall progressively further behind their peers in academic achievement and cognitive functioning. Fowler et al. (1988) found that older children with HbSS earned lower scores than younger children with HbSS on norm-referenced tests of reading, short term memory, and visual motor integration. Also, Wang et al. (2001) found that older children with HbSS with normal MRIs had lower Verbal IQ, math achievement, and processing speed than younger children with HbSS who also had normal MRIs. This is consistent with other studies that have found that younger children with SCD perform better than older children on tests of math achievement (Wasserman et al., 1991) and tests of visual-motor integration, attention and impulsivity (Brown et al., 1993). As for SES, only one study has examined it in relation to academic achievement in children with SCD. Devine, Brown, Lambert, Donegan, and Eckman, (1998) found that in a group of 74 youth with SCD aged 5-17 years, SES, as measured by parental education and income, was a strong predictor of academic achievement over and above the influence of illness parameters or family factors. Also, though not specific to children with SCD, low SES has been shown to negatively influence the academic achievement of healthy African American children (Brody, Stoneman, Flor, & McCrary, 1995). Overall, being older and having low SES are related to worse academic functioning in children with SCD.

Previous research has established that the academic performance of children with SCD is influenced by a number of factors, including neurocognitive functioning, disease severity, age, and SES. However, one factor that has not been extensively examined as a possible influencing

factor on academic outcomes of children with SCD is family functioning. Family functioning may directly influence academic achievement in youth with SCD, or may act as a protective factor in youth with SCD. The following section provides a review of the literature on family functioning of children with SCD and explores the research linking family functioning to academic outcomes in this population.

Family Functioning in Children with SCD

Family functioning can be defined as the environment created by the social patterns and the structural make-up of the family unit (Lewandowski, Palermo, Stinson, Handley, & Chambers, 2010). It describes the relationships between and among family members, as well as the context for their relationships. Elements of family functioning that are often studied include levels of adaptability, cohesion, conflict, organization, and communication. Well-functioning families are generally characterized as adaptable, cohesive, low in conflict, organized, and using clear and effective communication styles. In contrast, poorly functioning families splinter under stress and are characterized by disorganization or rigid control, poor communication, high conflict and poor affective regulation (Alderfer et al., 2008).

There are several frameworks that have been used to discuss family functioning in the literature, such as the Family Environment Model developed by Moos (1974). The Family Environment Model discusses family functioning by focusing on the climate and focal areas of a family. Moos and Moos (2009) characterize families on three dimensions: their relationships, their personal growth, and the systems maintenance of the family as a whole. The relationship dimension refers to the level of support and dedication within the family, and characterizes how cohesive, conflicted, and expressive family members are when interacting with each other. Personal growth refers to the extent to which family members are focused on and encourage the

self-development of each other in several different areas, including the area of achievement. System maintenance dimension refers to how important orderliness and structure are in planning events and setting rules, and characterizes how controlling and organized the family as a unit acts. Families that perform well in all of these areas are considered well-functioning according to the Family Environment Model, with the understanding that family functioning is on a continuum.

There have been mixed findings regarding whether families with children with SCD function differently than families without SCD. In a review of family functioning of families with children with SCD, Burlew, Evans and Oler (1989) reported that families with children with SCD demonstrate a lower level of family functioning than families with healthy children. Specifically, primary caregivers of children is SCD were found to be more stressed than parents of healthy same-aged children, secondary caregivers felt less like all their needs were met in the family than parents of healthy children, and healthy siblings' relationships with parents were found to be adversely affected. In another study of 78 families that had a preschooler with SCD and 72 families with healthy children (Evans et al., 1988), the same research group found that the families with children with SCD had elevated levels of conflict and control, and lower levels of organization. In contrast, Midence, McManus, Fuggle, and Davies's (1996) study of 39 families with children with SCD and 24 families with healthy children aged 6–16 years found that families with children with SCD were more cohesive than families with healthy children. Also, in a study using 77 children with SCD, 28 of their healthy siblings, and 74 youth not affected by SCD aged 5-18 years, Barbarin (1999) found that there were no differences between the families with children with SCD and families with healthy children matched on demographic factors. Thompson and his colleagues (1999) reported a similar finding for a sample of 289 children with

SCD aged 5 to 15 years. Specifically, they found that scores on the Family Environment Scale (FES) for families with children with SCD were not significantly different than the normative sample.

Of note, research conducted by Barbarin also by Thompson had much larger sample sizes than the work of Midence and the studies that made up the Burlew review. The research of Barbarin and Thompson also included a broader age range of participants than Evans' study. Additionally, Barbarin's (1999) study used a unique assessment device to assess family functioning, based on his previous research, which examined family relations, maturity demands and protectiveness. The other studies assessed family functioning with broadly used measures, such as the FES. These factors may account for the variability in findings. Taken together, findings indicate that families with school age children and adolescents with SCD do not appear to function differently than families with healthy children and adolescents (Barbarin, 1999; Devine et al., 1998; Noll et al., 1994; Thompson et al., 1999); however, families with young children with SCD, which are still learning to manage the disease, may experience lower levels of family functioning than families with healthy children (Brown et al., 2010).

Family Functioning and Academic Achievement in Children with SCD

Although the literature on the influence of family functioning on academic achievement in children with SCD is sparse, academic achievement has been firmly related to family functioning in populations other than youth with SCD. King's (1998) study of 346 college students found that FES scores were correlated with past high school performance and current college performance. Results indicated that high family conflict, low expressiveness, low cohesion, and low recreation orientation scores were related to low high school grade point averages (GPA). Additionally, high moral-religious orientation scores on the FES were related to

high GPAs in high school and good class attendance in college. In a study of rural southern African American families, Brody, Stoneman, Flor, and McCray (1995) collected data on 90 two-parent families whose oldest children were aged 9-12 years. They found that increased parental depression and family conflict reduced children' self-regulation and that lower self-regulation was associated with lower grades in reading and math. Thus, family functioning indirectly affected academic achievement via self-regulation. Lastly, in a review of the effects of family functioning on middle school students, Wentzel (1994) reported that parenting style, parent interaction and inter-parent hostility (conflict) all affect adolescents' self-regulation, and self-regulation is highly predictive of grade point average in middle school students.

A few studies have found that aspects of family functioning are related to academic functioning in youth with SCD. Barbarin (1994) integrated focus group data with case review and direct measures. Results indicated that youth with SCD were shown to have good academic, social and psychological outcomes when their parents had good mental health, when single parents had community support, and when their families established high expectations for the youth with SCD. Barbarin et al. (1999) also found that high parental expectations were predictive of high academic achievement in youth with SCD. Alternatively, findings from Devine and colleagues' (1998) study indicated that family functioning does not play a role in the academic functioning of students with SCD. These researchers studied 74 youth with SCD aged 5-17 years, and found that family functioning, as measured by the Family Adaptability and Cohesion Scale, second edition (FACES-II), did not predict academic achievement. The variation between Devine's results and Barbarin's results could be due to the different ways they measured family functioning. Barbarin's measure of family functioning assessed protectiveness over the sick child, amount of responsibility expected of the child, and the level of conflict in the family,

whereas Devine's measure assessed the families' the level of flexibility or rigidity, and the families' level of connectedness.

Family functioning may serve as a protective mechanism that modifies the risk of poor academic functioning in youth with SCD. Specifically, poorly functioning families may exacerbate the effects of the disease, while well-functioning families may ameliorate the risk of poor outcomes. Wentzel's (1994) research, previously discussed, describes a connection between family functioning and a child's self-regulation. Well-regulated children are able to set goals and boundaries for themselves better than unregulated children, and according to Wentzel's research, better regulated children also have higher GPA's than their unregulated peers. Research has established that the academic outcomes of children with SCD are influenced by variables such as high SCD severity, older age, and lower SES. These risk factors, however, do not hinder all children with SCD, as some show academic resiliency. Family functioning may lead to children with SCD developing more self-regulation, which may protect them against the negative effects of disease severity, age, and SES on their academic outcomes. Specifically, families with higher quality relationships, firm guidance and clear standards will have better regulated children, and better self-regulation may protect children with SCD against the academic risks associated with the disease. Similarly, families with poor relationships, guidance and standards will have children with poor self-regulation, which may lead to academic vulnerability in children with SCD. Good family functioning may reduce the effects of disease severity, age and SES on academic achievement, and poor family functioning may increase the effects of these variables. There is no existing literature on whether good family functioning acts as a protective factor on the relationship between risk factors and academic outcomes, thus more research is needed.

The Current Study

SCD has been shown to negatively affect academic achievement in children, and factors related to poor academic achievement in children with SCD include poor neurocognitive functioning due to stroke, high disease severity, older age, and low SES. Research is lacking, however, in the relationship of family functioning to academic achievement in children with SCD. In light of research indicating family functioning affects the academic performance of healthy students, and preliminary research linking aspects of family functioning to academic achievement in youth with SCD, the current study seeks to add to the literature in this area by describing the way that family functioning and academic achievement are associated in students with SCD. The primary aim of the current study is to examine whether family functioning is directly related to the academic outcomes of youth with SCD above and beyond other psychosocial and medical factors. It was hypothesized that strong family functioning (e.g., positive family relationships, good systems maintenance, and high achievement orientation) would have a positive direct impact on the achievement scores over and above the influence of neuropsychological functioning, disease severity, SES, and age. The secondary aim of the study is to investigate whether family functioning moderates the relationship between disease severity, SES, age, and academic outcomes. It was hypothesized that strong family functioning would weaken the influence of high disease severity, low SES, and older age on academic outcomes in children with SCD.

CHAPTER II: METHODS

Participants

The participants in this study were children with SCD participating in phase three of the Cooperative Study of Sickle Cell Disease (CSSCD; Biologic Specimen and Data Repository Information Coordinating Center, 2008; Gaston & Rosse, 1983). The CSSCD was a longitudinal study to track the clinical course of SCD in patients from birth to adulthood that lasted from 1978 to 1998. The CSSCD was sponsored by the National Heart Lung and Blood Institute and was conducted by the Division of Blood Diseases and Resources of the National Institutes of Health. Twenty- three sites participated in the first ten year phase of the CSSCD with 4,085 participants across four age-based cohorts: newborn, pediatric, adolescent, and adult. Phase two was a 5 year study that followed up with 467 participants from the pediatric and infant cohorts from phase one. Phase three followed the cohort of phase two to collect longitudinal data on factors affecting overall disease severity, brain abnormalities, pulmonary dysfunction, neurocognitive and psychosocial functioning (CSSCD, 2008), and lasted from 1994 to 1998. The current study used data collected at the beginning of phase three from a pool of 378 participants.

Procedures

A sample of children and adolescents between the ages of 6 to 16 years old was pulled from the beginning of phase three of the CSSCD database. Data on demographics, family functioning, academic functioning, disease severity, and cognitive functioning were extracted for each participant from the respective code books: Roster, Family Environment Scale (FES), Neuropsychological Data and Achenbach Child Behavior Checklist (CBCL), History, and Neuropsychological Data. Participants without complete measures of interest were excluded. The data were originally collected via interview and individual testing at the participant's clinic site.

Measures

Demographic information. Basic demographic information on all participants was collected from the roster and history code books of phase 2 and 3 of CSSCD data. Age and sex information is located in the roster code book. The patients' grade and household income are located in the history code book.

Academic Functioning. The Woodcock-Johnson Tests of Achievement, Revised (WJ-R Ach; Woodcock & Johnson, 1989) is a widely administered, norm referenced, standardized test of academic achievement. High scores indicate better achievement in academic subjects, including reading and math, which are measured by the Broad Reading cluster and the Broad Math cluster respectively. The Broad Reading Cluster score is based on word decoding and reading comprehension ability. The Broad Math Cluster score is based on the ability to solve computation and applied problems. The WJ-R Ach was shown to have acceptable concurrent and construct validity by comparing its subtests and results to other achievement tests including the Kaufman Test of Educational Achievement, the Peabody Individual Achievement Test, and the Wide Range Achievement Test-Revised (Woodcock & Johnson, 1990). The internal consistency (Chronbach's alpha) for each of the subtests falls between the high .80s and the low .90s, indicating that they are reliable measures.

The School Competence Scale of the Achenbach Child Behavior Checklist (CBCL; Achenbach & Edelbrock, 1983) is a 4 item, parent completed behavior rating scale for children that reflects the strength of a participant's school related behaviors. The school competence scale score is based on items that assess level of performance in academic subjects, grade retention, receipt of special education services, and school problems. The School Competence scale has a reported internal consistency of .57 to .64 across different sexes and ages of students

(Achenbach, 1991); however, the scale does demonstrate strong discriminant validity in that it can distinguish students having problems in school from normal samples (Achenbach & Edelbrock, 1979). Also, when compared to the School Social Behavior Scales (SSBS), there is strong evidence of convergent validity for the schools competence scale and social competence scale of the SSBS (Lowe, 1998). To overcome the low reliability of this measures, the school competence scale scores were converted to a dichotomous variable of competence in school with T scores less than and equal to 40 coded as low competence for the current study.

Family Functioning. The Family Environment Scale (FES; Moos & Moos, 2009) was used to measure family functioning. The FES was designed by Moos and Moos to measure the social climate of families. The FES is 90 questions and consists of ten subscales. Each item aligns with one subscale and each subscale is a part of one dimension. The instrument measures three dimensions of family environment: family relationship index (FRI), personal growth index (PGI), and system maintenance index (SMI). The current study used the FRI, the organization and control subscales of the SMI, and the achievement orientation subscale from the PGI. The FRI is made up of the cohesion, conflict, and expressiveness subscales. The SMI is made up of control and organization subscales. Achievement Orientation was the sole subscale used from the PGI, because it is believed to influence the outcome variable in this study, academic achievement. Internal consistencies are in an appropriate range for the FRI (.78), Organization (.75), Control (.67), and Achievement Orientation subscales (.64) (Alderfer et al., 2008; Moos & Moos, 2009). The FES has been found valid in several studies that compared it to other measures of family functioning, including the Family Assessment Device, the Family Adaptability and Cohesion Evaluation Scales, and Family System Test (Moos & Moos, 2009). In addition, FRI is considered a well-established measure for pediatric populations (Alderfer et al., 2008).

Disease Severity. Disease severity was determined by SCD genotype. The SCD genotype was gathered from the roster code book of Phase 2 and 3 of the CSSCD. HbSS and HbSβ° are the most severe types of SCD, while HbSC and HbSβ+ are milder forms of the disease (Redding-Lallinger & Knoll, 2006). HbSS and HbSβ° were grouped together and coded as 1 to indicate high disease severity. HbSC and HbSβ+ were grouped together and coded as 0 to indicate mild disease severity. Hemoglobin genotype is predictive of life expectancy and complications resulting from the disease (Platt, et al., 1994; Sebastiani et al., 2007).

Cognitive Functioning. The Wechsler Intelligence Scale for Children, Third Edition (WISC-III; Wechsler, 1991) was used as a control variable that represents each child's preexisting cognitive ability. The WISC -III is a norm referenced standardized intelligence test in the Wechsler family of tests. The WISC-III yields a Full Scale IQ (FSIQ), a Verbal IQ, and a Performance IQ. FSIQ measures general cognitive aptitude and served as this study's measure of cognitive functioning. Higher scores indicate higher cognitive ability. The WISC-III was a widely used intelligence test during the period the CSSCD was conducted. The FSIQ, which was used in the current study, is found to be both highly reliable (Chronbach's alpha .95; Wechsler, 1991) and has documented validity across several types including: convergent validity with the Peabody Picture Vocabulary Test—Revised (Carvajal, 1993), predictive validity as demonstrated with the Wechsler Individual Achievement Test (Weiss & Prifitera, 1995), and factor validity (Roid & Worrall, 1997).

Data Analysis

The data were stored in SAS datasets, but were managed in Excel and were analyzed using SAS and SPSS. Descriptive statistics on variables of interest were calculated. To evaluate the primary aim, multiple regression models were calculated predicting the broad reading and

math scores and a logistic regression model was calculated predicting school competence. Disease severity, SES, age, and family functioning variables (FRI, organization, control, achievement orientation) were used as predictor variables, and IQ and sex were used as control variables. The distributions of the dependent variables used in the multiple regressions, reading achievement and math achievement, were examined. The skew and the kurtosis of the original data set indicated that the variables were normally distributed. The skew and the kurtosis of each of the outcome variables indicated that the variables were non-normally distributed (reading achievement skew=-0.59, kurtosis =1.04; math achievement skew=-0.51, kurtosis =2.22; school competence skew=-0.81, kurtosis =-0.28). Thus, the inverse, the square root, and the natural log of the each of the dependent variable were calculated to determine whether transforming the variables would improve the distribution by making them more normal in form. None of these transformations were found to bring skew or kurtosis closer to 0, than the original dataset. Upon visual inspection each of the outcome variables was found to be unimodal; however, extreme observations were noted. Twelve observations were found to lay more than 2.5 standard deviations beyond the mean. Since these 12 observations were not found to significantly change the analysis, only the results calculated from the original version of the data set are presented.

To evaluate the secondary aim, the interactions between age, SES, and SCD type with the family functioning variables were included in the above mentioned model. Interactions terms were developed by first standardizing continuous predictor variables (age, SES, and family functioning variables) to reduce collinearity between the predictor variables and the interaction terms. Then SCD type and the standardized variables of age and SES were multiplied by each family functioning variable (FRI, organization, control, and achievement orientation) to create the interaction terms. Significant interactions, indicating the presence of a moderating

relationship, were identified by analyzing the individual significances of each interaction term's single degree of freedom *t* test, given that the omnibus *F* test of the model is significant. The presence of a moderator effect would be determined if the interactions between (a) age and family variables, (b) SES and family variables or (c) SCD type and family variables were found, while the main effects of age, SES, SCD type and family functioning were controlled. If an interaction term was found to be significant, simple slopes would be calculated for the relationship of the predictor variable to the outcome variable at three different levels (the mean, one standard deviation below the mean, and one standard deviation above the mean) of the moderating family functioning variable. If the simple slopes are not parallel, and significantly different than zero, this would demonstrate the moderating effect of the family functioning variable.

CHAPTER III: RESULTS

Descriptive Statistics

The overall sample extracted from the CSSCD dataset consisted of 198 children ranging in age from 6-16 years with a mean age of 10.30 (*SD*=2.72) and a mean grade level of 3.95 (*SD*=2.65, range=0-10). Descriptive statistics for the participants are listed in Tables 1 and 2. Of these participants, 91 were girls (45.96%) and 107 were boys (54.04%). One hundred ninety-three of the participants were black (97.47%) and 5 were another race (2.53%). One hundred twenty-eight participants (64.65%) had been diagnosed with HBSS, 59 (29.80%) with HBSC, 8 (4.04%) with HBSβ^{+}, and 3 (1.52%) with HBSβ°. The participants' families had a median household income between $10,000-14,999 and ranged from less than $5000 to $70,000-$99,999.

Table 1

Descriptive Statistics for Categorical Variables, (N=198)

	N	Percentage
Gender		
Male	107	54.04%
Female	91	45.96%
Race		
Black	193	97.47%
Other	5	2.53%
SCD Type		
HBSS	127	64.65%
HBSC	59	29.80%
HBSβ^+	8	4.04%
HBSβ°	3	1.52%
Income Level		
Less than $5,000	17	8.59%
$5,000-9,999	49	24.75%
$10,000-14,999	33	16.67%
$15,000-19,999	31	15.66%
$20,000-29,999	28	14.14%
$30,000-49,999	28	14.14%
$50,000-69,999	9	4.55%
$70,000-99,999	3	1.52%

Table 2

Descriptive Statistics for Continuous Variables, N=198

	Mean	**SD**	**Range**
Age	10.30	2.72	6-16
Grade	3.95	2.65	0-10
FSIQ[a]	82.13	14.34	40-129
Reading Achievement[a]	88.44	19.50	19-136
Math Achievement[a]	88.05	15.32	25-131
School Competence[b]	43.82	9.88	18-55
Family Relationship Index	168.93	22.20	91-209
Cohesion[b]	54.23	11.42	9-68
Conflict[b]	46.28	10.31	32-75
Expressiveness[b]	48.03	9.85	15-66
Organization[b]	57.08	10.34	26-70
Control[b]	58.74	7.70	32-76
Achievement Orientation[b]	54.84	7.05	35-72

Note. [a]reported as standard score (*M*=100, *SD*=15), [b]reported as T-score (*M*=50, *SD*=10)

FSIQ and achievement scores are reported as standard scores, which have a mean of 100 and a standard deviation of 15. The mean FSIQ of participants was 82.13 (*SD*=14.34, range=40-129), which is in the below average range. The mean reading achievement score was 88.44 (*SD*=19.50, range= 19-136), and the mean math achievement score was 88.05 (*SD*=15.32, range=25-131), both of which are in the average range. School competence scores were reported as T-scores, which have a mean of 50 and a standard deviation of 10. The mean school

competence score was 43.82 (*SD*=9.88, range=18-55), which is in the average range. The percentage of participants with a score of 40 or below, that is indicated to be at-risk for low school competences, was 34.1%. Participants' guardians completed the FES. Of those guardians, 173 were mothers of participants (88.72%), 6 were fathers (3.08%), and 16 had other relationships with the participants (8.21%). The FES subscales were reported as T-scores. The subscales that make up the FRI are cohesion (*M* = 54.23, *SD* = 11.42, range = 9-68), conflict (*M* = 46.28, *SD* = 11.42, range = 32-75), and expressiveness (*M* = 48.03, *SD* = 9.85, range = 15-66), all of which are in the average range. The mean FRI score was 168.93 (*SD* = 22.20, range = 91-209). The FES organization subscale score was 57.08 (*SD* = 10.34, range = 26-70), the FES control subscale score was 58.72 (*SD* = 7.69, range = 32-76), and the FES achievement orientation subscale score was 54.84 (*SD* = 7.05, range = 35-72), all of which are in the average range.

Correlations

Pearson product correlations were calculated for all of the variables used in the subsequent analysis, and can be found in Table 3. All of the outcomes variables (e.g., reading achievement, math achievement, and school competence) were positively correlated. Specifically, reading achievement was correlated to math achievement (*r* = .72) and school competence (*r* = .55) and math achievement was correlated to school competence (*r* = .55). Reading achievement was also significantly correlated with income (*r* = .36), sex (*r* = -.17), FSIQ (*r* = .70), FRI scores (*r* = .18), and FES control scores (*r* = -.14). Math achievement was significantly correlated with age (*r* = -.15), income (*r* = .32), FSIQ (*r* = .75), and FRI scores (r = .17). School competence was significantly correlated with age (*r* = -.17), FSIQ (*r* = .46), FRI scores (*r* = .28), and FES organization scores (*r* = .21). In addition, FRI scores were significantly

correlated with income (r = .16), FSIQ (r = .15), and FES organization scores (r = .45). FSIQ was also correlated with age (r = -.16) and income (r = .36). FES control scores were correlated to FES achievement orientation scores (r = .20).

Table 3

Correlations, N=198

		1	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**
1.	Age	-										
2.	Disease Severity	.06	-									
3.	Income	-.00	.06	-								
4.	Sex	-.10	.02	-.06	-							
5.	FSIQ	-.16*	.09	.36**	-.08	-						
6.	FRI	-.01	-.08	.15*	-.03	.15*	-					
7.	Organization	-.06	.02	-.13	.03	-.03	.45**	-				
8.	Control	-.07	.03	.02	.06	-.07	-.04	.07	-			
9.	Achievement Orientation	.01	-.01	.03	.06	-.06	.04	.00	.20**	-		
10.	Reading Achievement	.02	.05	.36**	-.17*	.70**	.18*	.03	-.14*	-.05	-	
11.	Math Achievement	-.15*	.13	.32**	-.11	.75**	.17*	.10	-.08	-.05	.72**	-
12.	School Competence	-.17*	.07	.06	.02	.46**	.28**	.21**	-.09	-.01	.55**	.55**

* p <.05, **p < .01

Regression Models Predicting Reading and Math Achievement

To test the primary aim, simultaneous regression models predicting reading and math achievement were calculated using age, sex, disease severity, income, FSIQ, FRI scores, FES organization scores, FES control scores, and FES achievement orientation scores (see Table 4 and 5). To test the secondary aim, the interactions between the family functioning variables (e.g., FRI, FES organization, FES control, and FES achievement orientation) and age, SCD type, and income were included in the previously described models.

The model for predicting reading achievement was significant and accounted for 50% of the variance (F (21, 176) = 10.38, $p < .01$). Reading achievement was significantly predicted by age and FSIQ. Specifically, age uniquely accounted for 1.38% of the variance ($\beta = .13$, $t = 2.33$, $p = .02$) and FSIQ uniquely accounted for 31.80% of the variance in reading achievement ($\beta = .65$, $t = 11.19$, $p < .01$). The model for predicting math achievement was significant and accounted for 55.62% of the variance (F (21,176) = 12.76, $p < .01$). Math achievement was significantly predicted by FSIQ, which accounted for 37.81% of the variance ($\beta = .71$, $t = 12.96$, $p < .01$). None of the family functioning variables or interactions added uniquely predicted reading or math achievement.

Table 4

Simultaneous Regression Analysis Predicting Reading Achievement, N = 198

	Reading Achievement				
	T	β	Partial R^2	F	Total Adjusted R^2
				10.38**	.50
Age	2.33	.13*	.01		
Disease Severity	.58	.03	.00		
Income	1.73	.10	.01		
Sex	-1.28	-.07	.00		
FSIQ	11.19	.65**	.32		
FRI	.53	.04	.00		
FES Org	-.36	-.03	.00		
FES Control	-1.65	-.10	.01		
FES AO	-.43	-.03	.00		
SCD Type × FRI	.38	.03	.00		
SCD Type × Org	.19	.01	.00		
SCD Type × Control	.42	.03	.00		
SCD Type × AO	.48	.03	.00		
SES × FRI	-.57	-.03	.00		
SES × Org	.19	.01	.00		
SES × Control	.28	.02	.01		
SES × AO	1.48	.08	.00		
Age × FRI	1.11	.07	.00		

Age × Org	-.05	-.00	.00
Age × Control	-.82	-.04	.00
Age × AO	-.30	-.02	.00

* p <.05, **p < .01

Table 5

Simultaneous Regression Analysis Predicting Math Achievement, N = 198

	Math Achievement				
	T	B	Partial R^2	F	Total Adjusted R^2
				12.76**	.56
Age	-.54	-.03	.00		
Disease Severity	1.56	.08	.01		
Income	1.24	.07	.00		
Sex	-.89	-.04	.00		
FSIQ	12.96	.71**	.38		
FRI	.10	.01	.00		
FES Org	1.02	.07	.00		
FES Control	-.55	-.03	.00		
FES AO	-.08	-.00	.00		
SCD Type × FRI	.82	.06	.00		
SCD Type × Org	-.46	-.03	.00		
SCD Type × Control	-.11	-.01	.00		
SCD Type × AO	.15	.01	.00		
SES × FRI	-.03	-.00	.00		
SES × Org	-1.02	-.06	.00		
SES× Control	1.16	.06	.00		
SES × AO	.99	.05	.00		
Age × FRI	.25	.01	.00		

Age × Org	-1.74	-.10	.01
Age × Control	-.80	-.04	.00
Age× AO	-1.15	-.06	.00

* p <.05, **p < .01

Logistic Regression Model Predicting School Competence

The school competence variable was dichotomized into T-scores over 40, indicating average or better school competence, and T-scores of 40 and under indicating at-risk or worse school competence. The model predicting school competence was analyzed using logistic regression and can be found in Table 6. The overall model was found to account for 22.5% of the variance *(-2LL* = 177.86, p = .002). Full scale IQ was significantly predictive of school competence (*OR* = 1.04, CI_{95} = 1.02, 1.06). In order to test the moderation hypothesis, interaction terms were included in the regression equation, but were not significant in the model. This indicates that family functioning does not buffer against the effects of disease severity, income, or age on school competence.

Table 6

Logistic Regression Model Predicting School Competence, N = 173

School Competence			
	β	OR (95% CI)	-2LL
			177.86**
Age	.07	1.07 (.72 – 1.60)	
Disease Severity	-.00	1.00 (.45 – 2.21)	
Income	-.30	.74 (.49 – 1.13)	
Sex	-.05	1.20 (.55 – 2.56)	
FSIQ	.04**	1.04 (1.02 – 1.06)	
FRI	.20	1.22 (.70 – 2.13)	
FES Org	.48	1.62 (.93 – 2.82)	
FES Control	-.42	.66 (.40 – 1.10)	
FES AO	-.05	.95 (.57 – 1.58)	
SCD Type × FRI	.41	1.50 (.61 – 3.68)	
SCD Type × Org	-.62	.54 (.23 – 1.28)	
SCD Type × Control	-.19	.83 (.31 – 2.22)	
SCD Type × AO	.521	1.68 (.70 – 4.05)	
SES × FRI	.31	1.37 (.88 – 2.13)	
SES × Org	-.14	.87 (.55 – 1.39)	
SES× Control	-.05	.96 (.60 – 1.52)	

SES × AO	.02	1.02 (.66 – 1.56)
Age × FRI	.14	1.15 (.74 – 1.81)
Age × Org	-.01	.99 (.64 – 1.53)
Age × Control	-.35	.71 (.44 – 1.15)
Age × AO	-.04	.97 (.64 – 1.45)

* p <.05, ** p < .01

CHAPTER IV: DISCUSSION

The primary aim of this study was to examine whether strong family functioning has a positive direct impact on academic outcomes of youth with SCD over and above other psychosocial and medical factors. Contrary to the hypotheses, results indicated that family functioning is not predictive of reading achievement, math achievement or school competence in children and adolescents with SCD over and above IQ, income, age and disease severity. IQ was the strongest predictor of reading and math achievement, and school competence when accounting for all other factors of interest. High IQ predicted high academic outcomes. Age was also a significant predictor of reading achievement, with older age being associated with higher reading achievement. This finding is in contrast to findings of Fowler and colleagues (1988) which found that older children with sickle cell anemia did worse on tests of reading than younger children, and the findings of Wasserman et al. (1991) and Wang et al. (2001), which observed that age was negatively related to math achievement and IQ, but not reading achievement. The findings of this study may be different than the previous findings because Fowler et al., Wasserman et al., and Wang et al. did not use IQ to predict academic achievement, but as one of their outcome variables. Findings did not account for the effects IQ had on reading achievement. The current study's findings indicate that if IQ were to be held constant across age, reading achievement would increase with older age. This implication is reasonable due to reading comprehension's heavy reliance on background knowledge, vocabulary, and practice, each of which should increase over time.

The secondary aim of the study was to investigate whether strong family functioning would weaken the influence of high disease severity, low SES, and older age on academic outcomes in children with SCD. Contrary to the hypotheses, family functioning did not buffer

against the negative effects of SCD severity, income or age on the academic outcomes of children with SCD. This result was unexpected considering the impact that family functioning has been shown to make on behavior (Thompson et al., 1999), mental health (Barbarin, 1994; Barbarin et al., 1999; Kell et al., 1998; Kliewer & Lewis, 1995), health care utilization (Barakat et al., 2007; Mitchell et al., 2007) and overall level of disability (Barakat et al., 2005; Graff et al., 2010) seen in children and adolescents with SCD. Family functioning may not have been found to be a buffer against risk factors related to poor academic achievement in children with SCD because IQ was such a powerful predictor of achievement that it overpowered any effects family functioning might have on achievement. Another reason family functioning may not have been found to buffer against poor achievement risk factors is family functioning's weak relationship to academic achievement. How well a family functions appears to have more direct relationships with other outcomes, such as how often families visit the emergency room and children's behavior, than academic achievement. Families are directly involved in behavior related to academic outcomes, such as close academic supervision and setting high academic expectations, but are not actually in control of the child's school performance. Families might also expend more resources on the child's physical wellbeing, and have fewer resources to devote to academic pursuits, which would mean that although a family may be well-functioning, their focus is not on academic outcomes. Overall, the findings of this study indicate that family functioning may not be the most effective point of intervention for the academic outcomes of children with SCD.

In this study, IQ was used to indicate neurocognitive functioning. IQ was the strongest predictor of academic achievement and school competence over and above all other variables, indicating that neurocognitive functioning is probably the most important predictor of school

outcomes of children with SCD. Of note, the mean IQ of the sample was below average. The likely explanation for this is that low mean IQ is an accurate representation of the neurocognitive functioning of the population of children with SCD. Past research has shown that when controlling for demographic characteristics, children with SCD are more likely than other children to have low IQ (Boulet, Yanni, Creary, & Olney, 2010) and that more children with SCD have special education placements than other African American children (Peterson et al., 2005). Wang et al. (2001) found similarly low mean neurocognitive scores in children with SCD who suffered from overt and silent strokes. The literature indicates that stroke is a major factor in neurocognitive functioning (Kral et al., 2006), but older age, lower hematocrit levels, hypoxia, and anemia (Armstrong, 2005) are also related to poor cognitive functioning. These findings may indicate that many of the children in the current study's sample have suffered from stroke, low hematocrit level, hypoxia or anemia, which would affect them neurocognitively.

Limitations

There are several limitations to the current study that should be considered when interpreting the results of this research. The greatest limitations are related to the use of preexisting data. Although the use of data from the CSSCD does greatly improve the ability to generalize the findings of this research due to its large sample size, several limitations are associated with the sample. First, there are missing data throughout the dataset. The current study removed participants that did not have all the variables of interests. By removing these participants, there may have been a specific group of participants that were not included, which would limit the generalizability of the findings. Another limitation is that the measures of school competence and family functioning rely entirely on parental report. In research on behavior and relationships, findings are considered more valid if they are supported across multiple

informants, such as parents, children and their teachers. Additionally, the CSSCD dataset does not contain a control group to compare the sample with SCD against, which would allow the study to detect differences between predictors of academic outcomes in children with SCD and healthy children.

Another limitation associated with using a preexisting dataset is the conceptual validity of the measures available through the dataset to answer the current research questions. For instance, IQ, which was used as an indicator of neurocognitive functioning, may not be the best indicator of neurocognitive functioning, but was the best available option to account for the construct. Other research has used neurocognitive testing batteries, such as the Luria-Nebraska Neuropsychological Battery (Wasserman et al., 1991), which are less dependent on the crystalized knowledge and verbal abilities that are highly predictive of school performance, but more predictive of the declines in cognitive proficiency associated with SCD. Also, SCD genotype was used as the sole measurement of disease severity because it was the most feasible method for measuring disease severity. This method is not as comprehensive, however, as other measurements of disease severity, which include using sickled hemoglobin concentration, frequency and duration of pain episodes (Dampier et al., 2010), number of SCD related complications, days of hospitalization (Eaton et al., 1995), and composites using various combinations of these variables (Mayes et al., 2011). By using SCD genotype as the only measure of disease severity, this research does not consider all the levels of variation that exist in the range of disease severity. Lastly, household income, which was grouped into 6 categories, was used as the measure of SES, but this measure does not take into account the number of people contributing to the household, or the number of dependents in the household. Having the per capita household income would have taken into account the number of people the household

income supported. Per capita household income is often used as an indicator in SES in psychological research (Brody et al., 1998). Household income also does not take the participants' parents' occupation or education level. Using a composite index of these factors, such as the Hollingshead four-factor index (Hollingshead, 1975), may provide a more valid estimate of SES. The Hollingshead index is a commonly used measure of SES in pediatric SCD research (Fowler et al., 1988; Burlew et al., 1989; Kell et al., 1998; Gold, Mahrer, Treadwell, Weissman, & Vichinsky, 2008).

A fourth limitation of the current study is that family environment, as measured by the FES, is not the only conceptualization of family functioning. Family environment, and particularly the family relationship portion of family environment, is a reliable and valid measure of family functioning that has been validated for the pediatric SCD population (Alderfer et al., 2008); however it does not conceptually address all areas of family functioning that may be related to academic achievement. There are several frameworks that are used to describe family functioning in the literature, such as the Circumplex Model (Olsen, 2011), which looks at balance in cohesion flexibility and communication, the Beavers System Model (Beavers & Hampson, 2000), which looks at family competence and style, and the McMaster Model (Epstein, Baldwin & Bishop, 1983) which focuses on the relation dyads within the family and how they solve problems. These components are all important factors in family adaptation that the family environment model does not consider. Neither does the family environment model take parental mental health into consideration, which has been shown to directly affect the psychosocial outcomes of children with SCD (Barbarin, 1999; Edwards et al., 2006; Tunde-Ayinmode, 2007).

Clinical Implications and Future Directions

The findings of the current study suggest possible clinical implications and draw attention to topics and methods to be used in future research. The greatest clinical implication of this research is that IQ seems to be the pivotal factor predicting the academic outcomes of children with SCD, and that these children have lower IQs than similar children without SCD. This finding is consistent with the previous research (Brown et al., 1993; Fowler et al., 1988; Swift et al., 1989; Wang et al., 2001, Wasserman et al., 1991). For example, Wang et al. (2001) found that children with SCD that were classified in groups with lower IQ scores had lower achievement scores than children with SCD in groups with higher IQ scores. In Wasserman et al.'s 1991 study, children with SCD had lower mean IQ and achievement scores than their healthy siblings. Overt and silent stroke are contributing factors to these low IQ scores, along with low hematocrit levels, anemia and hypoxia. Prevention of stroke and the detection of physiological factors related to poor neurocognitive performance are high priorities in improving the academic outcomes of children with SCD. Transcranial Doppler Ultrasonography has been found to be a viable imaging method for detecting neurological abnormalities that predict stroke and poor neurocognitive functioning (Kral et al. 2006). Also, in a 1998 clinical trial, Adams et al. found that children with SCD at risk for stroke who received blood transfusions were less likely to suffer a stroke than those that did not receive transfusion. According to Gulbis and colleagues (2005), hydroxyurea is a promising treatment for the prevention of stroke in children with SCD and can be used with children under the age of two years old. Mallick and Ganesan (2008) recommend the use of blood transfusion, hydroxyurea, and/or annual transcranial Doppler ultrasounds for the primary prevention of stroke in this population.

Early academic intervention and monitoring also seem to be important for children with SCD. Research has shown that higher mental and physical functioning before a stroke is predictive of better functioning after stroke (Nys et al., 2005). In an effort to learn as much as possible before neurocognitive abilities begin to decline, early educational intervention, such as Head Start, could be used to promote early literacy and numeracy. Research has shown that preschool aged children with SCD usually have average intelligence, but low school readiness (Chua-Lim, Moore, McCleary, Shah, & Mankad, 1993). Early intervention programs like Head Start are designed to improve school readiness. Tarazi, Grant, Ely, and Barakat (2007) found that children with SCD who attended preschool had better language skills than those that did not. Children with SCD should also receive regular neuropsychological testing so that their progress or regression can be more clearly detected. Frequent monitoring of neuropsychological functioning would help improve the type and timeliness of intervention, which may help children with SCD improve their achievement and keep up with their peers. In addition, children with SCD that are found to show poor neurocognitive functioning may benefit from cognitive intervention strategies that help improve specific domains of neurocognitive functioning. For example, if evaluation reveals poor executive functioning, than the child may benefit from direct instruction in how to formulate and execute plans, and frequent reminders of plans, such as electronic planners and visual schedules. If sustained attention is found to be poor, a child may benefit from being given short directions, and having frequent breaks during class to ensure efficient use of the child's attentional abilities. If visuo-spatial abilities are shown to be poor, the child may benefit from receiving occupational therapy.

Lastly, further research in all factors that contribute to academic resiliency in children with SCD is needed. Self-regulation has been shown to contribute to healthy children's academic

success (Wentzel, 1994), and active coping has been shown to contribute to the social-emotional health of children with SCD (Kliewer & Lewis, 1995). Research investigating possible linkages of factors such as these to academic functioning in children and adolescents with SCD could provide new avenues for intervention in the future.

Conclusion

The current study's findings contribute to the sparse literature centering on social and emotional factors contributions to academic outcomes in children and adolescents with SCD, specifically family functioning's relation to academic outcomes. Research establishing the role the family plays in the relationships between academic achievement, school functioning, and SCD is needed to better understand the way the social environment may affect children with SCD. While this study did not find a relationship between family functioning and academic outcomes, further research in the area is needed to make a definitive statement on the relationship between the two. Cognitive functioning, as indicated by IQ, was the strongest predictor in the current study of reading achievement, math achievement, and school competence in children with SCD. Cognitive functioning should be a point of intervention for the academic outcomes of children with SCD, and should be further investigated. As the life span of people with SCD continues to increase, more research is needed on improving factors, like academic outcomes, that contribute to their overall success in life.

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APPENDIX

IRB Approval

EAST CAROLINA UNIVERSITY
University & Medical Center Institutional Review Board Office
4N-70 Brody Medical Sciences Building· Mail Stop 682
600 Moye Boulevard · Greenville, NC 27834
Office 252-744-2914 · Fax 252-744-2284 · www.ecu.edu/irb

Notification of Continuing Review Approval: Expedited

From: Biomedical IRB
To: Cecelia Valrie
CC:

Date: 8/21/2012
Re: CR00000524
UMCIRB 08-0660
[IMPORTED] The Influence of Neuropsychological Functioning on Adjustment in Children and Adolescents with Sickle Cell Disease

The continuing review of your expedited study was approved. Approval of the study and any consent form(s) is for the period of 8/21/2012 to 8/20/2013. This research study is eligible for review under expedited category #7. The Chairperson (or designee) deemed this study no more than minimal risk.
Changes to this approved research may not be initiated without UMCIRB review except when necessary to eliminate an apparent immediate hazard to the participant. All unanticipated problems involving risks to participants and others must be promptly reported to the UMCIRB. The investigator must submit a continuing review/closure application to the UMCIRB prior to the date of study expiration. The Investigator must adhere to all reporting requirements for this study.

The approval includes the following items:

Name	Description	Modified	Version
Consent | History	Consent Forms	8/30/2011 8:58 PM	0.01
Memo from NHLBI | History	Additional Items	8/30/2011 9:05 PM	0.01
NHLBI Data Distribution Agreement | History	Additional Items	8/30/2011 9:05 PM	0.01
Protocol | History	Study Protocol or Grant Application	8/30/2011 7:31 PM	0.01

The Chairperson (or designee) does not have a potential for conflict of interest on this study.

IRB00000705 East Carolina U IRB #1 (Biomedical) IORG0000418
IRB00003781 East Carolina U IRB #2 (Behavioral/SS) IORG0000418 IRB00004073