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Predictors of Academic Achievement for School Age Children with Sickle Cell Disease

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Abstract

BACKGROUND—Children with sickle cell disease (SCD) are at risk for neurocognitive impairment and poor academic achievement, although there is limited research on factors predicting academic achievement in this population. This study explores the relative contribution to academic achievement of a comprehensive set of factors, such as environmental (socioeconomic status), disease-related (stroke, transfusion therapy, adherence), and psychosocial variables (child behavior, child quality of life (QoL)), controlling for intellectual functioning (IQ).

METHODS—Eighty-two children with SCD completed measures assessing IQ and academic achievement, while parents completed questionnaires assessing adherence, child behavior, and child quality of life. Medical chart reviews were conducted to determine disease-related factors.

RESULTS—Hierarchical regression analyses indicated that 55% of the variance in academic skills was accounted for by IQ, parent education, chronic transfusion status, and QoL [$R^2 = .55$, $F(5, 77) = 18.34$, $p < .01$]. Follow-up analyses for broad reading [$R^2 = .52$, $F(5, 77) = 16.37$, $p < .01$], and math calculation [$R^2 = .44$, $F(5, 77) = 12.14$, $p < .01$] were also significant.

CONCLUSION—The findings suggest a significant contribution of factors beyond IQ to academic achievement. Findings allow for identification of children with SCD at risk for academic difficulties for whom psychoeducational interventions may enhance academic achievement.

Keywords

Chronic Diseases; Children with Disabilities; School Health Services; School Psychology

Sickle cell disease (SCD) is a genetic and chronic health condition that occurs in one in every 400 African American births and one in every 1,200 Hispanic American births (National Heart, Lung, and Blood Institute [NHLBI], 2008). This disease primarily affects individuals of African descent, but is also found in individuals from South or Central America, the Caribbean islands, Mediterranean countries, India and Saudi Arabia (Noll et al., 2001; Smith, 1999). SCD is inherited through one sickle hemoglobin gene from each parent or one sickle hemoglobin gene from one parent and another abnormal hemoglobin gene from the other parent (Brawley et al., 2008). The hallmark complication of sickle cell disease is painful vaso-occlusive episodes, which cause acute pain due to blocked blood flow or chronic pain caused by damage from repeated ischemic episodes (Mousa et al., 2010; Yale, Nagib, & Guthrie, 2000). Other significant medical complications include chronic anemia, elevated risk of infection, stroke, acute chest syndrome, splenic sequestration, retinal injury, kidney damage, and delayed growth (Brown, Buchanan, & Doecke, 1993).

Children with the most common form of sickle cell disease, sickle cell anemia or HbSS, experience more frequent and severe symptoms than children with other forms of the disease such as HbSC (Austin, Cohen, & Losseff, 2007). For example, children with HbSS disease are at highest risk of experiencing stroke (Kral et al., 2001), which affects approximately 1% of children with HbSC and HbS β^+ thalassemia and approximately 5% of children with HbSS (Ohene-Frempong et al., 1998;). Stroke in youth with SCD represents one of the most serious complications due to its potential deleterious impact on neurocognitive functioning (Brown, Armstrong, & Eckman, 1993). However, even children with SCD without evidence of stroke (i.e., on neuroimaging) demonstrate neurocognitive impairment relative to their healthy peers (Nettles, 1994; Noll et al., 2001; Steen et al., 2005).

For example, children with SCD demonstrate specific deficits in attention, concentration, reading decoding (Brown, Buchanan, et al., 1993), and executive skills (Schatz & Roberts, 2007). Furthermore, children with SCD score lower than their healthy counterparts on measures of intelligence (Schatz, Finke, Kellett, & Kramer, 2002), specifically in the areas of crystallized ability, processing speed, and short-term memory (Schatz, Finke, & Roberts, 2004). There is also evidence that children with SCD demonstrate increased incidence of school absences, repeating grades, and special education services relative to their healthy peers, suggesting a potential interaction between neurocognitive, environmental and disease variables (Dyson et al., 2010; Fowler, Johnson, & Atkinson, 1985).

The heightened risk for problems in academic achievement may be due to the individual, combined, and/or interactive effects of the disease, psychosocial factors, and environmental variables on cognitive functioning (Brown, Armstrong, et al., 1993; Schatz, Finke, & Roberts, 2004). For example, chronic anemia directly influences neurocognitive functioning through decreased oxygen supply (Schatz et al., 2006) and may indirectly affect children with SCD through associated fatigue (While & Mullen, 2004), which can result in difficulty paying attention in school. In addition, children with SCD with neurological dysfunction also demonstrate increased behavioral problems in school related to impaired executive function and are therefore more likely to be retained or receive special education services (Schatz, 2004).

In terms of environmental variables, research has documented the negative effects of low socioeconomic status (SES) or family income on neurocognitive functioning above and beyond the contribution of disease factors in children with SCD (Tarazi, Grant, Ely & Barakat, 2007), many of whom come from lower SES backgrounds (Brown, Armstrong, & Eckman, 1993). In contrast, for those children who come from families with higher SES, environmental factors may protect against the direct and indirect effects of the disease on academic achievement (Brown, Buchanan, et al., 1993). In addition to IQ and SES, parent education is positively correlated with academic achievement in children with SCD (Fowler et al., 1988) and may be the most critical aspect of SES, as economically disadvantaged children or children with less educated parents may have fewer opportunities for learning and stimulation (Brown, Armstrong, et al., 1993; Davis-Kean, 2005).

Additionally, children with SCD demonstrate an increased risk for behavioral problems (Lamanek, Moore, Gresham, Williamson, & Kelley, 1986; Thompson et al., 1999) and internalizing disorders (Yang 1994). Moreover, these youth are at risk for reduced health-related quality of life (HRQoL)(Barakat, Patterson, Daniel, & Dampier, 2008), although many children with SCD nonetheless demonstrate adaptive coping skills, including interpreting minor health events without increased anxiety (Fuggle, Shand, Gill & Davies, 1996). A review by Anie (2005) suggests that better adjustment to SCD may be due to various psychosocial factors including lower perceived stress, more adaptive family functioning, and reduced negative coping, such as catastrophizing, and passive coping, such as passive adherence to typical medical recommendations regardless of the nature of the pain crisis (Gil, Abrams, Phillips, & Keefe, 1989). While there is some research linking psychosocial factors to neurocognitive or academic achievement in children with SCD (Tarazi, et al., 2007), more evidence is needed to better understand the contribution of these variables.

The extant literature is equivocal regarding risk and resilience factors for academic achievement in children with SCD (Richard & Burlew, 1997). Armstrong and Horn's neurodevelopmental model of chronic illness posits that neurocognitive functioning in children with a chronic illness is the result of an interaction between age at diagnosis and the time at which functioning is assessed, extent of disease treatment or severity, successful treatment adherence, time since stroke, and non-disease factors such as environmental influences (Armstrong, 2006). Using this model, we conceptualized risk for problems with academic achievement in children with SCD in terms of environmental factors, disease-related factors, and psychosocial factors. In contrast to prior studies (Brown, Buchanan, et al., 1993; Schatz, 2004), this study sought to examine a more comprehensive set of factors that may influence academic achievement for children with SCD, which could enable school personnel to identify those at highest risk for poorer academic achievement and design and implement targeted prevention and intervention efforts.

We hypothesized that environmental variables (parent education, income), disease-related factors (average hemoglobin, genotype, stroke, chronic transfusion, adherence, nutrition) and psychosocial variables (child behavior and QoL) would be significantly associated with academic achievement above and beyond the contribution of IQ. In addition, we separately examined associations between the same set of predictors (environmental variables, disease-related factors, and psychosocial variables) on math and reading abilities.

Methods

Participants

The study sample was drawn from baseline participants in a randomized controlled study examining the efficacy of a problem-solving intervention for families with a school-age

child with SCD. Participants were eligible for the study if they had a diagnosis of SCD, were between the ages of 6 and 12 years, were primarily English speaking and received follow-up care at one of two children's hospitals in a northeastern city in the United States. Exclusion criteria included the presence of a developmental disability or severe psychopathology in either caregivers or children that would negatively affect their ability to participate in the problem-solving intervention. Children with less severe behavioral or developmental concerns (e.g., attention-deficit/hyperactivity disorder, anxiety disorder, reading problems) were retained in the study.

Of the initial study sample, 82 participants (50% female) consented and were enrolled in the study. Children ranged in age from 6 to 12 years ($M = 8.42$, $SD = 2.10$) and were in kindergarten to eighth grade ($Mdn = 3$, $SD = 2.18$). Participants primarily identified as African American and Non-Hispanic (94%), with the majority of the additional families identifying as Caribbean or South/Central American. The majority of children had SS disease (61%), while the remaining children had SC disease (28%), sickle β^+ thalassemia (7%), sickle β^0 thalassemia (1%), SO Arab (1%) and SJ Baltimore (1%). Sixteen percent of the sample ($n = 13$) had a history of stroke (overt and/or silent infarct) according to medical chart review. Most caregivers (94% female, $M = 37.79$ years of age) had completed either some college or vocational school (40%). See Table 1 for further descriptive characteristics of the sample.

Instruments

The *General Information Form*, completed by the primary caregiver, assessed basic demographic information (age, gender, ethnicity, education, self-reported income within response options of \$0 - \$5,000 with increments of \$833 (refer to Table 2) of family member participants.

Disease Factors—Disease factors including genotype, average hemoglobin (past three blood draws), and history of chronic transfusion treatment (noted by physician) were determined through medical chart reviews. In addition, lifetime occurrence of stroke was noted if there was any mention of overt or silent stroke in the medical chart. Research assistants systematically documented complications, disease information, and healthcare utilizations in the year prior to participating in the study. Genotype was dichotomized into more severe types (i.e., HbSS and HbS β^0) and less severe types (i.e., HbSC and HbS β^+), a practice which has been used in other studies of neurocognitive functioning in children with SCD (Schatz & Roberts, 2007). Although there are exceptions, children with more severe genotypes typically demonstrate increased SCD complications including stroke and greater health care utilization, while children with less severe genotypes typically demonstrate fewer neurocognitive and medical complications (Austin, Cohen, & Losseff, 2007; Charache, Lubin, & Reid, 1989).

The *Self Care Inventory-Sickle Cell* (SCI-SC; (Hilker, Jordan, Jensen, Elkin, & Iyer, 2006) is a parent report questionnaire that measures level of adherence in SCD care behaviors. The SCI-SC consists of 19 items, 18 of which assess self-care behaviors and one that assesses amount of sleep. The parent rates how well their child follows doctor's instructions for their SCD care on a 5-point Likert-type scale (1 = never to 5 = always). Subscales for the SCI-SC include General Health Behavior, Sickle Cell Management, and Pain Management. For these analyses, a total score was used rather than the General Health Behavior score, as it encompasses all three factors and a separate nutrition score (computed from two items specifically assessing nutrition). The addition of the nutrition score was chosen in order to isolate the unique contribution of this construct, considering problems with nutrition which children with SCD experience (Brown, Armstrong, et al., 1993; Finan et al., 1988;). Higher

scores on this measure indicate better medical adherence. The SCI-SC is considered a reliable and valid measure of adherence in SCD care behaviors (Hilker et al., 2006). For this sample, nutrition $\alpha = .88$ while total score $\alpha = .77$.

Psychosocial Factors—Parents completed the *Child Behavior Checklist* (CBCL; Achenbach, 1966), a measure consisting of 140 items assessing internalizing and externalizing symptoms in children and adolescents using a 3-point Likert-type scale (0 = not true to 2 = very true). The Total Behavior Problems score was used to evaluate the relationship between child behavior and academic achievement for this study. The CBCL is a reliable and valid measure of child behavior and had an α of .82 for this sample (Achenbach, 1966).

The *Pediatric Quality of Life Inventory* (PedsQL; Varni et al., 1998) is a 23-item parent form which assesses a child's quality of life on a 5-point Likert-type scale (0 = never to 4 = almost always). The PedsQL consists of four scales: Physical Functioning, Emotional Functioning, Social functioning, and School Functioning and yields three summary scores including physical health, psychosocial health (emotional, social and school functioning), and a total score. Higher scores on this instrument indicate better quality of life. The Total Score was used for this study. This measure has been validated in healthy and chronically ill populations (James W. Varni, Seid, & Kurtin, 2001), and had an $\alpha = .91$ for this sample.

The Hematology/Oncology Psycho-Educational Needs Assessment (Peterson, Palermo, Swift, Beebe, & Drotar, 2005) consists of 17 items assessing areas of academic/school functioning, including learning problems, functional impairment, behavioral concerns at school, and evaluation of current services. Items were developed by a multidisciplinary SCD team. Items used in this study include questions addressing school absences (days absent in past year), presence of an individualized education plan (IEP) or 504 Plan (yes/no), and classroom accommodations (number of accommodations provided by teacher).

Cognitive Functioning and Academic Achievement—The *Wechsler Abbreviated Scale of Intelligence* (WASI; Ryan & Brown, 2005) is a brief measure of intelligence for individuals ages 6 to 89 years. The WASI can be administered in its two-subtest or four-subtest form. The two-subtest form, which was used for this study, can be administered in 15 minutes and includes Vocabulary and Matrix Reasoning subtests. The WASI is a reliable and valid measure of intelligence (Canivez, Konold, Collins, & Wilson, 2009; Ryan & Brown, 2005).

The *Woodcock-Johnson-III* (WJ-III) is a measure of academic achievement for individuals ages 2 through 90 (Grenwelge, 2009). The WJ-III was validated for use in the United States, with norms included by grade. Reliability measures indicate Cronbach alpha's of 0.80 or higher (Grenwelge, 2009). Subsets of the WJ-III Tests of Achievement were used for the study, including Calculation, Spelling, Letter-Word Identification, Reading Fluency, Math Fluency and Passage Comprehension. Academic skills (Letter-Word Identification, Calculation, Spelling), Broad Reading (Letter-Word Identification, Reading Fluency, Passage Comprehension), and Math Calculation Skills (Calculation, Math Fluency) clusters were used for analyses. The Math Calculation Skills and Broad Reading Clusters were selected for further analyses, as these skills are most commonly screened in children with SCD (Brown et al., 2000; Puffer, Schatz, & Roberts, 2010; Schatz et al., 2001). The WJ-III Tests of Achievement have been used in previous studies to measure academic functioning (Schatz, 2004) and are considered a standard measure of academic achievement for children with SCD (Daly, Kral, & Tarazi, 2011).

Procedure

Participants and caregivers were recruited during outpatient clinic visits, inpatient hospital stays, and SCD community events between 2009 and 2012. Assent was obtained from child participants and permission/consent was obtained from parents. Assessments were conducted in the family's home or in an outpatient medical setting during clinic visits, with a typical duration of two hours. The WJ-III Tests of Achievement were administered first and took approximately 45 - 60 minutes to complete, with the remainder of the paper-and-pencil measures taking 60 minutes. A sticker map was used to maintain attention and breaks were given as needed. The study protocol was approved by the Human Subjects Committees of the appropriate Institutional Review Boards.

Data Analysis

Preliminary correlations (Pearson and Spearman's correlations as appropriate) were computed between potential predictors and the academic skills cluster of the WJ-III (Letter Word Identification, Calculation, Spelling), which provides an overall score of basic achievement skills. The assumptions of linearity, independence, homoscedasticity, and normality were met for each regression conducted. Missing income data (in instances where caregivers chose not to disclose) and summary WJ-III scores (for children unable to generate score) were replaced by the mean of the variable. For follow-up, exploratory analyses correlations were computed with the broad reading cluster (Letter Word Identification, Reading Fluency, and Passage Comprehension) and the math calculation cluster of the WJ-III (Math Fluency and Calculation) to determine differences in predictors of math and reading skills. Variables significant in Pearson or Spearman correlations at $p < 0.05$ were entered into hierarchical regressions. Order of entry was determined by controlling for IQ in the initial steps, followed by environmental factors, disease factors, and finally psychosocial factors. Based on power estimates, a sample size of 50 and above is deemed adequate for regression analysis (Wilson Van Voorhis & Morgan, 2007).

Results

Preliminary Analyses

Overall, participants scored within the average range compared to their same-age peers on the WJ-III subtests measuring general academic skills ($M = 96.40$, $SD = 16.42$), broad reading ($M = 93.28$, $SD = 15.21$), and math calculation ($M = 92.40$, $SD = 13.78$). Intellectual functioning, based on WASI 2-subtest Full Scale IQ scores, also fell within the average range compared to same aged peers ($M = 93.62$, $SD = 12.14$). PedsQL total scores in our sample reflected below average quality of life compared to other children with chronic illness ($M = 70.49$, $SD = 16.42$) (Dampier et al., 2010). Mean total scores on the SCI ($M = 11.39$, $SD = 1.94$) and SCI nutrition subscale ($M = 3.87$, $SD = .98$), which reflect treatment adherence behaviors, fell within the average range compared to other children with chronic illness. The mean average hemoglobin of our sample is consistent with that seen in other samples of children with SCD ($M = 9.46$, $SD = 1.55$) (Simon, 2009). More detailed results of the preliminary analyses are presented in Table 2.

Results of bivariate correlations, computed to determine which variables would be included in the regression analysis, are presented in Table 3. SCI-SC total score, average hemoglobin, SCI-SC nutrition, total school absences, and CBCL total behavior problems were not significantly correlated with WJ-III subscales, and thus were not included in subsequent analyses. T-tests revealed that performance on academic skills was not associated with gender ($M_{\text{girls}} = 99.28$, $M_{\text{boys}} = 93.45$, $t(79) = 1.61$, $p = .11$).

Regression Analyses

Hierarchical regressions were used to assess the contribution of each relevant variable to the prediction of academic achievement. For each regression analysis, variables were entered in the following order: (1) WASI total score; (2) Parent education; (3) SCD disease-related factors including chronic transfusion and lifetime occurrence of stroke (medical chart review); and, (4) Psychosocial factors (PedsQL total score). The regression model for the academic skills cluster, presented in Table 4, explained 55% of the variance [$R^2 = .55$, $F(5, 77) = 18.34$, $p < .01$]. As expected, IQ, parent education, chronic transfusion and QoL emerged as significant predictors of academic achievement. Specifically, children with higher scores on the WASI, more highly educated parents, with better quality of life, and those not on chronic transfusion scored higher on the academic skills cluster of the WJ-III.

In order to examine variation in predictors across math and reading skills, follow-up analyses were conducted. For broad reading and math calculation regression analyses, WASI total score, parent education, disease-related variables, and psychosocial factors were entered in the same order as the academic skills regression. The regression model for broad reading (see Table 5) was significant and accounted for 52% of the variance in academic achievement [$R^2 = .52$, $F(5, 77) = 16.37$, $p < .01$]. The regression model for math calculation (see Table 6) also was significant and accounted for 44% of the variance [$R^2 = .44$, $F(5, 77) = 12.14$, $p < .01$]. These results indicate that children with higher scores on the WASI, more highly educated parents, better quality of life, and those not on chronic transfusion scored higher on the math calculation cluster of the WJ-III. Alternatively, children with higher scores on the WASI, more highly educated parents, and better quality of life scored higher on the broad reading cluster.

Discussion

Children with SCD are confronted with disease, environmental, and psychosocial challenges that can impact academic achievement. Disease complications for these children include neurocognitive impairment due to chronic anemia and/or stroke, difficulty participating in class due to pain or fatigue, and health-related symptoms that can result in school absence. Complicating psychosocial and environmental factors include increased risk of reduced quality of life, limited access to education and economic disadvantage. Despite many potential risk factors, predictors of academic achievement have not been well studied in this population. The purpose of this study was to examine the relative contribution of a range of disease, environmental, and psychosocial factors to academic achievement in children with SCD.

Previous studies have found lower scores in children with SCD compared to same aged peers on measures of academic achievement (Schatz, Finke, Kellett, & Kramer, 2002). The mean scores of youth in the current sample, however, fell within the average range compared to normative sample means on measures of academic skills, math calculation, and broad reading. As such, despite the myriad of factors that place children with SCD at risk for compromised educational outcomes, the current sample demonstrated resilience in terms of performance on academic achievement tests. One possible explanation for this finding is that children in this sample benefitted from the care provided by a comprehensive sickle cell center that is equipped to educate and guide school personnel to support children with SCD in the classroom. For example, in a study examining the efficacy of a school intervention program, teachers who had received the education intervention demonstrated increased knowledge about SCD, including the ability to identify signs of stroke and understanding of the importance of hydration (Koontz, Short, Kalinyak, & Noll, 2004).

Parent education was a significant predictor of academic achievement, a finding which is consistent with investigations of children without chronic health conditions (Supplee, Shaw, Hailstones, & Hartman, 2004). Interestingly, family income was not correlated with academic achievement in this study, suggesting that parent education may be a more salient factor for identifying children with SCD who may benefit from early identification, monitoring, and/or intervention to promote academic development. Parent education has been indirectly associated with academic achievement in an African-American sample through beliefs about their child's achievement and responsiveness and stimulation of the family environment (Davis-Kean, 2005) and to neurocognitive functioning in children with SCD (Tarazi et al., 2007). However, to our knowledge, the current study is the first to identify the association between parent education and academic achievement in a pediatric SCD population. Although most of the families in this sample live below the poverty line, over half of the caregivers attended at least some college, reflecting national trends that point to lower income for African Americans with the same level of education compared to Whites (Aud, Fox, & KewalRamani, 2010). It is likely that more highly educated caregivers are equipped to assist their children with strategies to succeed in school despite the risk of low SES, for example by monitoring homework assignments and communicating with the child's school. Therefore, interventions that encourage parental participation may be able to assist less educated parents in developing strategies to support their child's education as well as convey beliefs surrounding the importance of education and the benefit of academic effort in obtaining future opportunities.

Disease-related factors including average hemoglobin, genotype, nutrition, and adherence were not significantly associated with academic achievement. In contrast, chronic transfusion status emerged as a significant predictor of academic achievement such that children on chronic transfusion treatment scored lower than those children not receiving transfusion treatment. Chronic blood transfusions are a common treatment for children who have suffered a stroke or who are at risk for a stroke (Routhieaux, Sarcone, & Stegenga, 2005), suggesting that lower scores in the chronic transfusion group may reflect the direct, negative effects of SCD complications on neurological integrity and, in turn, cognitive functioning and academic achievement. In addition, the frequency of transfusion treatment (every 3 to 6 weeks) increases school absences and could thereby contribute to poor academic achievement (Routhieaux et al., 2005). Although stroke status did not emerge as a significant independent predictor of academic achievement in this sample, many previous studies provide evidence for an association between stroke status and cognitive and academic achievement for children with SCD (Camargo de Oliveira, Ciasca, & Moura-Ribero, 2008; Harriman et al., 1991). Due to the small group of children with a history stroke in the current study, it is possible that there was insufficient power to detect an association between stroke status and academic achievement.

Less is known about the impact of psychosocial factors on academic achievement in children with SCD. What is known is that children with SCD demonstrate more limitations across all QoL domains compared to healthy children from similar SES backgrounds, including physical health, psychological functioning, and social functioning (Palermo, Schwartz, Drotar, & McGowan, 2002). In this sample, parent-reported child QoL was a significant predictor of academic achievement, with children who demonstrate better QoL scoring higher on measures of academic achievement. The relationship between QoL and academic achievement may be bidirectional, with academic achievement predicting QoL in some cases. Alternatively, it is possible that an overall better QoL may allow children to engage consistently in their education, resulting in improved school performance. Also, children with better QoL may have learned to more effectively manage physical problems (e.g., mild pain, fatigue) in a classroom setting and may use social resources to offset frequent absences by asking a classmate to collect missed assignments. In addition, children

who report higher QoL may be engaging in response shift, changing one's conceptualization of QoL and thereby appraising their health status as more favorable (Sprangers & Schwartz, 1999), which could contribute to more adaptive functioning in the classroom despite pain and fatigue. The results of this study indicate that adaptive physical, social, emotional, and school functioning may positively affect academic achievement or, alternatively, that better academic achievement contributes to increased QoL. In contrast to previous research that demonstrates a relationship between behavior problems and deficits in academic achievement (Thompson et al., 1999), child behavior problems were not correlated with academic achievement in this sample, suggesting that behavioral problems did not significantly interfere with academic achievement.

Additional analyses for math calculation and broad reading clusters of the WJ-III generally revealed similar predictors of academic achievement across the two skills areas. Because specific neurocognitive deficits in children with SCD most often occur in the areas of attention and executive function rather than a particular subject (Brown, Buchanan, et al., 1993; Brown et al., 2000; Jeffrey Schatz et al., 2002), a similar set of predictors for each subject area is expected. One notable exception was chronic transfusion treatment, which predicted scores on the math calculation cluster but not on the broad reading cluster of the WJ-III. The presence of chronic transfusion as a predictor suggests that the adverse effects of receiving chronic transfusion, such as school absences, may have a more detrimental effect on the development of math skills as compared to reading skills. Additionally, children on chronic transfusion treatment typically demonstrate more severe effects of the disease, likely impacting academic achievement.

Limitations

One limitation of this study is that academic achievement was assessed at only one time point. According to the neurodevelopmental model, the extent of cognitive impairment depends on the time at which the disease or treatment impacts the central nervous system (CNS) and the time at which the child is assessed (Armstrong, 2006). Future research should consider cognitive impairments over time by assessing children at multiple time points. In addition, longitudinal study designs can account for the contribution of chronic anemia and changes in SCD severity to poor functioning on the WJ-III for children on chronic transfusion. Additional tests of attention and executive functions would also strengthen the methodology of future research studies, given the pervasiveness of these specific deficits in children who demonstrate neurocognitive impairment. Of note is the low incidence of stroke in our sample compared to the general SCD population, which could have contributed to the overall average scores on the WASI and WJ-III (Ohene-Frempong et al., 1998; Schatz, Brown, Pascual, Hsu, & DeBaun, 2001). Children who have experienced a silent stroke but did not have transcranial Doppler ultrasonography (TCD) or magnetic resonance imaging (MRI) in the past year could not be accounted for in our sample.

Another limitation is that the sample was drawn largely from an urban dwelling population, possibly limiting the ability to generalize the results to other regions. Although parent ratings of child QoL and behavior were obtained, future research incorporating ratings from children and teachers would enable a more comprehensive investigation of the factors that impact academic achievement. In addition, understanding how child, parent, and teacher ratings differ might assist in developing interventions that more specifically target problems in their systemic context.

Implications for Practice

Due to our findings that most of the sample scored within the average range on measures of academic achievement, the majority of children with SCD may require only minor

interventions from school personnel in order to optimize learning. Academic functioning is optimized when parents, school administrators and teachers, physicians, and mental health professionals work together and are educated on the cognitive outcomes of SCD as well as the laws protecting children with chronic health conditions in school (Daly, Kral, & Brown, 2008). Along with modifications at home, teachers that are willing to send assignments home, explain missed assignments, and in some cases provide alternative assignments can promote optimal school functioning for children with SCD (Lightfoot, Wright, & Sloper, 1999). In addition, teachers and school-based mental health professionals can support students with chronic illnesses in their relationships with peers and manage minor pain episodes by working with the school nurse to allow children access to medications when needed (Dyson, Atkin, Culley, & Dyson, 2007).

It is important to note that a subset of our sample was performing below age expectations. These findings are consistent with the literature and suggest that some children with SCD may be struggling academically, but are not receiving accommodations and special education services that would likely allow them to reach their full potential in the classroom. For example, although almost 16% of the sample had experienced stroke, only about 25% of the sample had an IEP or 504 plan. This low percentage likely reflects the children with the most severe deficits that had already been noted by teachers and/or parents prior to the beginning of the study. Early identification and treatment of children at risk for stroke as well as increased support for parents in obtaining special education services for their child may be instrumental in further supporting academic development in children with SCD (Burchinal & Campbell, 1997). Because of the risks to general cognitive development, children with SCD may require learning accommodations, such as early, small group instruction, close monitoring of progress, and/or accommodations in the classroom to promote optimal development of academic skills. School mental health professionals have a critical role in advocating for children with SCD in the IEP process as well as in collaborating with teachers and special education teams, modeling positive communication, highlighting student strengths, and increasing family involvement (Geltner, 2008).

Our findings reveal that there are disease factors in addition to stroke that influence academic achievement (e.g., chronic transfusion) and psychosocial factors such as quality of life play an important role in determining those at risk. Consequently, in addition to frequent screening for neurocognitive deficits (Daly, Kral, & Tarazi, 2011), a comprehensive screening program that identifies children with SCD who demonstrate additional environmental and psychosocial risk factors may aid in detecting a broader range of children at risk for poor academic achievement. A more comprehensive understanding of the factors that influence academic achievement in children with SCD will allow professionals to best target their efforts toward serving these children.

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Table 1

Descriptive Characteristics of the Sample

Demographic and Medical Variables (N = 82)	n (%)	Mean (SD)
Age (6- 12yrs)		8.43 (2.09)
Gender (Female)	41 (50.0)	
Grade (K-8)		3.02 (2.17)
Ethnicity, Non-Hispanic	77 (93.9)	
Lifetime Occurrence of Stroke/CVA	13 (15.9)	
Accommodations in Classroom	52 (63.4)	
IEP	12 (14.6)	
504 Plan	11 (13.9)	
Genotype		
HbSS	50 (61.0)	
HbSC	23 (28.0)	
Hb Beta ⁺ Thalassemia	6 (7.3)	
Hb Beta ⁰ Thalassemia	1 (1.2)	
SO Arab	1 (1.2)	
SJ Baltimore	1 (1.2)	
Average Hemoglobin		9.48 (1.55)
On Chronic Transfusion	19 (23.2)	

Table 2

Academic Functioning and Predictors

Variables (N=82)	n	(%)	Mean	SD	Range
Woodcock Johnson Cluster Scores					
Academic Skills			96.44	16.33	45 – 129
Broad Reading			93.37	15.13	45 – 125
Math Calculation			92.38	13.70	55 – 124
WASI IQ Score			93.61	12.07	65 – 121
Monthly Income					
\$0 – \$833	5	(6.1)			
\$834 – \$1,666		21	(25.6)		
\$1,667 – \$2,499	15	(18.3)			
\$2,500 – \$3,333	23	(28.0)			
\$3,334 – \$4,166	3	(3.7)			
\$4,167 – \$4,999	5	(6.1)			
Over \$5,000	10	(12.2)			
Highest Level of Education (Primary Caregiver)					
Some High School	9	(11.0)			
High School Graduate	21	(25.6)			
Some College/Vocational School/Associates Degree	33	(40.2)			
Bachelor's Degree	12	(14.6)			
Professional School/Graduate School	7	(8.5)			
SCI Total Score			11.40	1.93	7.67 – 15.00
SCI Nutrition			3.87	.98	1 – 5
Days Absent Last Year					
1 – 7 Days	40	(48.8)			
8 – 14 Days	25	(30.5)			
15 – 20 Days	9	(11.0)			
20+ Days	8	(9.8)			
CBCL Total Problems			25.59	20.42	1 – 113
PedsQL Total Score			70.10	17.24	33.70 – 96.74

Table 3

Correlations

Variable	1	2	3	4	5	6	7	8	9	10	11	12	13
1. WJIII Academic Skills	–	.53**	.21*	.51**	.12	-.04	-.27*	-.28*	.09	.10	-.11	-.14	.32**
2. WASI IQ Score	.53**	–	.22	.29**	.11	.03	-.06	-.05	.08	.12	.17	-.25*	.07
3. Income	.21	.22	–	.48**	.08	.18	-.10	-.26*	.08	.13	–	-.17	.21
4. Parent Education	.51**	.29**	.48**	–	-.01	-.15	-.09	-.08	-.02	-.08	-.11	-.12	.12
5. Genotype	.11	.11	.08	-.01	–	.42**	-.13	-.22*	-.05	.02	-.18	.11	.19
6. Average Hemoglobin	-.04	.03	.18	-.15	.42**	–	-.21	-.28*	.13	.05	-.04	.17	.07
7. Stroke	-.27*	-.06	-.10	-.09	-.13	-.21	–	.47**	-.06	-.13	-.02	-.08	.06
8. Chronic Transfusion	-.28*	-.05	-.26*	-.08	-.22*	-.28*	.47**	–	-.03	-.08	.18	-.12	.03
9. SCI Total Score	.09	.08	.08	-.02	-.05	.13	-.06	-.03	–	.56**	-.04	–	.33**
10. SCI Nutrition	.10	.12	.13	-.08	.02	.05	-.13	-.08	.56**	–	-.09	-.24*	.18
11. Total Absences	-.11	.17	–	-.11	-.18	-.04	-.02	.18	-.04	-.09	–	.09	–
12. CBCL Total Problems	-.14	-.25*	-.17	-.12	.11	.12	-.08	-.12	-.44**	-.24*	.09	–	-.31**
13. PedsQL Total Score	.32**	.07	.21	.12	-.19	.07	.06	.03	.33**	.18	–	–	–
											.32**	.31**	

* p<.05,

** p<.01

Table 4

Academic Skills Regression Analysis

	β	t	$F(df)$	R^2
Full Model			18.34 (4,81) ***	.55
Final Step				
IQ	.39	4.86 ***		
Parent Education	.33	4.09 ***		
Stroke	-.14	-1.63		
Chronic Transfusion	-.17	-1.99 *		
PedsQL Total Score	.26	3.34 ***		

*
p<0.10,

**
p<.05,

p<.01

Table 5

Broad Reading Regression Analysis

	β	t	$F(df)$	R^2
Full Model			16.37 (4,81) ***	.52
Final Step				
IQ	.42	5.09 ***		
Parent Education	.31	3.70 ***		
Stroke	-.15	-1.63		
Chronic Transfusion	-.14	-1.56		
PedsQL Total Score	.22	2.76 ***		

*
p<0.10,

**
p<.05,

p<.01

Table 6**Math Calculation Regression Analysis**

	β	t	$F(df)$	R^2
Full Model			12.14 (4,81) ***	.44
Final Step				
IQ	.33	3.67 ***		
Parent Education	.29	3.22 ***		
Stroke	-.14	-1.46		
Chronic Transfusion	-.20	-2.05 **		
PedsQL Total Score	.24	2.78 ***		

*
p<0.10,

**
p<.05,

p<.01