

ESTABLISHING VALIDITY FOR MEASURES OF EXECUTIVE FUNCTIONING AND BEHAVIOR IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE

by

Lee Nathaniel Purvis

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Director of Thesis: Cecelia Valrie, Ph.D.

Major Department: Psychology

This study examined the criterion validity of the Behavior Rating Inventory for Executive Functioning (BRIEF; Gioia, Isquith, Guy, & Kenworthy, 2000) and the Behavior Assessment System for Children, 2nd Edition (BASC-2; Reynold & Kamphaus, 2004) for children and adolescents with sickle cell disease (SCD) in a clinically referred pediatric SCD population. It was predicted that criterion validity would be supported for the BRIEF and BASC-2 as evidenced by the participants from the clinically referred sample having elevated levels of executive dysfunction on the BRIEF's Behavior Regulation Index and Metacognitive Index as compared to the normative sample of the BRIEF, and elevated levels of behavioral and emotional problems and lowered adaptive functioning on the BASC-2's internalizing, externalizing, and adaptive domains as compared to the normative sample of the BASC-2. It was also predicted that criterion validity would be supported for the BRIEF and BASC-2 as evidenced by the participants from the clinically referred sample with SCD and stroke evidencing more executive dysfunction and behavioral problems as compared to those with SCD and without stroke. Comparisons of mean scores were made between pediatric SCD patients and the criterion T-score of 50 and between pediatric SCD patients with and without stroke. The percentage of participants' ratings in the at-risk and clinical ranges on the BRIEF and

BASC-2 were compared to 16%, which is the standard percentage of the normative sample within these classifications and they were also compared to the percentage of participants with SCD and without stroke. Results from t-test analyses indicated that the mean of the participants' T-scores on the BRI and MI of the BRIEF and externalizing, internalizing, and adaptive scales BASC-2 were significantly different than the criterion T-score of 50 in every comparison. No significant differences were found with regards to stroke status. Results from chi-square analyses indicated a higher percent of participants with SCD fell in the at-risk and clinical ranges on the BRIEF and BASC-2 when compared to 16% in every comparison but not in regards to stroke status. These results suggest that the BRIEF and BASC-2 can detect deficits in executive functioning and behavior among children and adolescents with sickle cell disease but more research is needed to determine if these measures can detect differences between participants with SCD with and without stroke.

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BEHAVIOR IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE

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by

Lee Nathaniel Purvis

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Lee Nathaniel Purvis

APPROVED BY:

DIRECTOR OF THESIS:

Cecelia Valrie, Ph.D.

COMMITTEE MEMBER:

Christy Walcott, Ph.D.

COMMITTEE MEMBER:

Jennifer Kazmerski, Ph.D.

CHAIR, DEPARTMENT OF PSYCHOLOGY:

Susan L. McCammon, Ph.D., CIP

DEAN OF THE GRADUATE SCHOOL:

Paul J. Gemperline, Ph.D.

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CHAPTER 1: INTRODUCTION

Children and adolescents with sickle cell disease (SCD) are at risk for behavior problems (Thompson et al., 2003), and executive dysfunction, such as deficits in attention and memory (Bonner, Gustafson, Schumacher, & Thompson, 1999). However, it is difficult to assess these types of problems in children with SCD due to a lack of measures that have been validated with this population. Identification of behavioral or executive functioning concerns in the school system is necessary to ensure proper accommodations are in place to address these concerns. The following sections include a description of SCD, criterion validity, and a discussion of two measures that may be useful for accurately evaluating behavior problems and executive functioning in children and adolescents with SCD.

Description of Sickle Cell Disease

SCD is an autosomal recessive genetic disorder that results in the production of abnormal hemoglobin (Schatz & McClellan, 2006). This abnormal hemoglobin, HbS, results in red blood cells forming an abnormal crescent shape that obstruct normal blood flow, resulting in anemia and blockages (Bonner et al., 1999). These blockages lead to vaso-occlusive crises (VOCs), or recurring episodes of pain. SCD occurs in approximately 2,000 births annually in the United States and in about 1 out of 365 African American births (Daly, Kral, & Tarazi, 2011). There is also an increased frequency of SCD in descendants of individuals from Turkey, Greece, southern Mediterranean areas, Saudi Arabia, India, the Caribbean, and Latin America (Lemanek & Ranalli, 2009). The most common and severe SCD genotype is HbSS, which comprises two-thirds of all cases (Steinberg, 1984). Other SCD genotypes include HbSC, HbS beta thalassemia-plus, and HbS beta thalassemia-zero, which appear less frequently (Angastiniotis & Modell, 1998). Children and adolescents with HbSS and HbS beta thalassemia-zero tend to exhibit increased medical complications due to SCD than the other genotypes. The mean life expectancy

is 42 years for males with HbSS, 48 for females with HbSS, 60 years for males with HbSC, and 68 years for females with HbSC (Platt et al., 1994).

Complications of SCD

Stroke. One medical complication of SCD is stroke, which is seen in approximately one-fourth to one-third of all individuals with SCD (Schatz & McClellan, 2006). There are two types of stroke: overt and silent. Overt strokes are strokes that can be seen with magnetic resonance imaging (MRI) and are accompanied by behavioral indicators, such as loss of speech or problems with motor control. Overt strokes occur in approximately five to eight percent of the HbSS population (Schatz & McClellan, 2006). In contrast, silent strokes are strokes that, despite being detectable with MRI, are not accompanied by behavioral indicators as seen in overt strokes (Schatz & Puffer, 2006). Thus, they are particularly hard to detect, as MRIs are not standard clinical practice and a silent stroke can occur anytime without overt behavioral symptoms. Silent strokes occur in approximately 22-35% of children with SCD before age 14 (Miller et al., 2001). There is a 70% recurrence rate of silent and overt stroke over a three-year period if no form of blood transfusion therapy has been performed (Schatz & Puffer, 2006). Both overt stroke and silent stroke are serious complications of SCD due to the impairments that individuals with SCD may develop in relation to attention, memory, visual-spatial processing, and academic achievement (Peterson, Palermo, Swift, Beebe, & Drotar, 2005).

Executive Functioning. A common complication associated with SCD is deficits in executive functioning. *Executive functioning* is the system or cognitive processes that regulate our deliberate control of thoughts, emotions, and behaviors, such as planning, working memory, initiation of tasks, attention, inhibition, and problem solving (Pennington & Ozonoff, 1996).

In order to investigate the relationship between neuropsychological performance and neuroimaging data in children with SCD, Berkelhammer and colleagues (2007) conducted an extensive review of the literature. Twenty-eight studies in the pediatric SCD literature published between 1991 and 2005 were reviewed with median sample sizes of 30 children with SCD. Forty three percent of the studies included a control group comprised of either siblings without SCD, healthy controls, or test norms. The researchers found that children with SCD and stroke experienced greater deficits in executive functioning in regards to working memory and attention than those children with SCD and without signs of stroke. This finding is consistent with results from Schatz, Brown, Pascual, Hsu, and DeBaun's (2001) study of 19 children with HbSS and silent stroke, 18 of their healthy siblings, and 45 children with SCD and without silent strokes aged 8-15 years examining educational outcomes and the rate of neuropsychological deficits associated with silent infarction. The study found that children with SCD and silent stroke were more likely to evidence poor sustained attention and executive functioning than healthy siblings and children with SCD and without signs of silent stroke. Nabors and Freymuth's (2002) study of 12 children with HbSS and stroke, 14 children with HbSS and without stroke, and 13 healthy siblings aged 6-13 years found that children with SCD and stroke had slower response times on tests of intelligence than the other two groups. They also noted that studies examining children with SCD that included no MRI information should be interpreted with caution because children with SCD may have had undocumented strokes.

In addition, deficits in executive functioning may present themselves in children and adolescents with SCD and without stroke. Brown and colleagues (1993) studied 70 children with SCD and without stroke aged 2-17 years and 18 siblings without SCD aged 3-15 years. The

study found that children with SCD and without stroke experienced deficits in sustained attention compared to sibling controls.

Notably, delays in cognitive functioning based on traditional IQ scores may be a result of executive dysfunction because these tests measure, among other constructs, some elements of executive functioning (e.g., working memory and sustained attention). And, there have been several studies that have shown that children and adolescents with SCD score lower on overall IQ when compared to healthy controls (Berkelhammer et al., 2007; Schatz & McClellan, 2006; Steen et al., 2005). Steen and colleagues' (2005) study of 54 children with HbSS and 54 healthy children with an average age of 11 years found that cognitive deficits can still be seen in children with HbSS and no evidence of stroke. Specifically, they found that 30 of the 54 patients with HbSS and a normal MRI scored lower on the Wechsler Intelligence Scale for Children Revised (Wechsler, 1991) and Wechsler Intelligence Scale for Children, 3rd Edition (Wechsler, 1974) by 12.9 points when compared to healthy controls matched on socioeconomic status and education level, which supported previous research findings indicating that even without evidence of stroke, the disease itself is associated with severe cognitive impairment (Steen et al., 2005).

Behavioral Problems. Behavioral problems can consist of externalizing, internalizing, and adaptive problems (Armstrong et al., 1996). *Externalizing problems* are disruptive behaviors such as poor control of temper, hyperactivity, or aggression (Gaultney, Merchant, & Gingras, 2009). *Internalizing problems* are self-directed negative behavior or mood states, such as depression and anxiety (Gaultney et al., 2009). *Adaptive* functioning encompasses three broad domains: conceptual, social, and practical and these three domains impact how well people function and respond to situations or events in their everyday lives (American Association of Mental Retardation, 2002).

To examine internalizing and externalizing behavior problems in children with SCD, Hijmans and colleagues (2009) investigated scores from the Child Behavior Checklist (CBCL; Achenbach, 1991), the Teacher Report Form (TRF; Achenbach, 1991), and the Disruptive Behavior Disorder rating scale (DBD; Pelham, Gnagy, Greenslade, & Milich, 1992). For children with SCD, these norm-referenced measures allow for the comparison of scores to same age peers to determine significance levels. Questionnaires were returned for 106 children aged 6-19 years with SCD and 37 healthy siblings, matched for age and sex. Results indicated that children with SCD have significantly higher mean scores on Internalizing Problems, which include Anxious/Depressed and Somatic Complaints, than healthy siblings and the norm population. They also found that those with more severe SCD genotypes were rated by caregivers as having higher scores on the Total Problems and Internalizing Problems scales. Of note, these findings are consistent with results from Rodrigue, Streisand, Banko, and Kedar's (1996) study of 85 children with SCD aged 4-8 years and 46 healthy controls. The parents of the children with SCD rated their children on the CBCL as higher on the Internalizing Problems scale compared to parents of the healthy children. Markedly, teachers rated children with SCD on the Teacher Report Form (TRF) as exhibiting less externalizing problems than their healthy peers. Noll, Reiter-Purtill, Vannatta, Gerhardt, and Short's (2007) study of 39 children with SCD aged 8-15 years and 38 healthy peers found that teachers rated children with SCD as being more prosocial and less aggressive than their healthy peers. No children with SCD were rated in the clinical range for internalizing problems. These findings were consistent with a longitudinal study by Noll, Kiska, Reiter-Purtill, Gerhardt, and Vannatta (2010) of 60 children with SCD and 66 healthy controls aged 10-17 years which found that children with SCD were perceived by peers and teachers as less aggressive. Brown, Eckman, Baldwin, and Buchanan's (1995) study of

61 African American children with SCD with an average age of nine years examined internalizing and externalizing problems. The study found only two children rated in the clinically significant range for internalizing problems and only one child for externalizing problems (T-score >70) on the CBCL. Thus, previous research suggests internalizing problems appear to be a more significant problem for children and adolescents with SCD than externalizing problems as noted by teachers and parents. However, few children and adolescents with SCD tend to fall into the clinical ranges for internalizing or externalizing problems.

As for adaptive functioning, findings indicate that children and adolescents with SCD experience problems in social and academic contexts. Hijmans et al. (2009) had teachers rate children with SCD and healthy siblings on the TRF of the Achenbach System of Empirically Based Assessment (ASEBA). Teachers scored children with SCD lower on Total Adaptive Functioning, and its subscales, Academic Performance and Behaving Appropriately, than healthy siblings. Children and adolescents with SCD who exhibit maladaptive behaviors were also noted to have poorer school performance that often leads to academic retention. Peterson and colleagues (2005) found that a higher number of adolescents with SCD are suspended and retained in comparison to children without SCD based on 72 parent reports. Also, Noll and colleagues' (2007) study indicated that peers rated children with SCD as having fewer friends and less athletic skills.

Rating Scales for Executive and Behavioral Functioning

The CBCL and TRF, two measures from the ASEBA, are the two most commonly used behavior problem checklists used with children and adolescents with SCD and provide assessments of frequently presenting symptoms in children and adolescents (Drotar, Stein, & Perrin, 1995). A limitation with the CBCL and TRF noted by Drotar et al. (1995) is that the

measures lack sensitivity to detect differences in symptoms that are within the normal range. Although children and adolescents with SCD have problems with executive and behavioral functioning, they tend not to be rated in the clinically significant range; however changes in symptoms in the average range may still impact their ability to adapt to their illness.

In order to better understand the utility of behavior and executive functioning rating scales for mild symptom changes within normal limits as it applies to SCD, a review of the validity of the measures is needed. The following sections describe validity and how it is assessed and then presents two measures, the Behavior Rating Inventory for Executive Functioning (BRIEF; Gioia, Isquith, Guy, & Kenworthy, 2000) and the Behavior Assessment System for Children, 2nd Edition (BASC-2; Reynold & Kamphaus, 2004), that may be of use for working with children and adolescents with SCD.

Description of Validity. *Validity* is obtained by determining the degree to which a measure assesses the variable it claims to assess (Gravetter & Forzano, 2006). There are several types of validity, such as predictive, criterion, concurrent, construct, and convergent. The current study focuses on criterion validity. *Criterion validity* is established for a measure when it accurately measures the event that is being observed (Kaplan, Bush, & Berry, 1976).

BRIEF. The BRIEF is a questionnaire designed to assess difficulties in executive functioning in school-age children and adolescents in an ecologically valid manner (Gioia et al., 2000). The authors of the BRIEF provide both a parent and teacher form. The BRIEF takes approximately the same amount of time to administer as other rating scales, such as the Conners Rating Scale-R (Conners, 1997), but has been used in evaluating children with more neuropsychologically based conditions, such as Tourette's syndrome (Mahone et al., 2002), traumatic brain injury (Mangeot, Armstrong, Colvin, Yeates, & Taylor, 2010), and autism (Gilotty, Kenworthy, Sirian, Black, &

Wagner, 2002). The BRIEF has also been noted as an appropriate measure to assess executive functioning because of its strengths in identifying the day-to-day behavioral impact of stroke in children (Gioia et al., 2000). This strength makes the BRIEF particularly useful for children and adolescents with SCD as this population is at a higher risk for stroke. The BRIEF may be able to act as an alarm for the possibility of stroke if the child's composite scores indicate clinical significance that were previously not a concern. Also, it is an inexpensive measure that requires less time to administer when assessing impairments in executive functioning than a full neuropsychological battery.

The majority of studies that have used the BRIEF use this measure to assess executive functioning in children with ADHD (e.g., Toplak, Bucciarelli, Jain, & Tannock, 2009; McAuley, Chen, Goos, Schacher, & Crosbie, 2010). Toplak and colleagues' (2009) study of 46 adolescents with a confirmed clinical diagnosis of ADHD and 44 adolescents without a diagnosis of ADHD aged 13-18 years compared the BRIEF to performance-based measures of executive functioning (e.g., the Stop Task, Trail Making Test, and the Stockings of Cambridge task). The study found that parent and teacher ratings of the BRIEF were better predictors of ADHD status than performance-based measures despite modest correlations. A study by McAuley et al. (2010) found that the two indices of the BRIEF, which include the Behavior Rating Index (BRI) and the Metacognition Index (MI), were found to significantly correlate with parent and teacher ratings of ADHD symptoms, impairment, and youth's level of academic proficiency. Toplak et al. (2009) also reported that the BRIEF's two indices are efficient in assessing global behaviors of executive functioning and the separate scales are efficient in assessing more narrow-band behaviors of executive functioning, such as organizing learning materials and the ability to initiate tasks. The BRIEF is not commonly used in the school setting, but by examining it in

children and adolescents with SCD, it will hopefully become a tool that can be used more frequently in the schools and particularly for kids living with a chronic illness, or who have possibly had a stroke.

BASC-2. The BASC-2 is a questionnaire designed to assess behavioral and emotional problems as well as adaptive functioning in children and adolescents using parent-, teacher-, and self-report forms (Reynolds & Kamphaus, 2004). The current study will only investigate the Parent Report Scale (PRS) and the Teacher Report Scale (TRS). The BASC-2 provides a quick overview of parents' and teachers' perceptions of a child's behavior which can then be used to tailor interventions in the area where the child is rated highly in, such as hyperactivity or aggression. Notably, Wolfe-Christensen, Mullins, Stinnett, Carpentier, & Fedele (2009) indicated that the BASC-2 is comparable to the CBCL in that they both assess and evaluate an array of behavior profiles, such as ADHD, learning disabilities, speech and hearing deficits, and developmental delay that can aid in making differential diagnoses in children. However, the BASC-2 also has composites that focus specifically on school and learning problems. Hijmans et al. (2009) listed better measurement of adaptive, executive functioning, and attention as additional advantages of the BASC-2 over the CBCL.

Studies That Use the BRIEF or BASC-2 to Assess Children and Adolescents with SCD

Research evaluating both the BRIEF and BASC-2 with children with SCD is limited. Kral and Brown (2004) have conducted the only study using these measures. They investigated the association between Transcranial Doppler (TCD)-measured cerebral blood flow velocity and various behavioral problems using the BRIEF and the BASC-2. The study consisted of 62 children and adolescents with SCD between the ages of 6 years and 16 years with no documented history of stroke, traumatic brain injury (TBI), birth complications, or other chronic

illness. All participants had the HbSS subtype of SCD. The groups were differentiated by their TCD status. The three categories of participants were normal TCD, conditional TCD, and abnormal TCD. The first edition of the BASC was completed by parents and teachers to assess internalizing, externalizing, and adaptive behaviors and the BRIEF was completed by parents and teachers to assess everyday behaviors associated with executive dysfunction in the home and school environments. Kral and Brown compared each TCD group and anticipated elevated levels of behavioral problems and executive dysfunction in groups with more atypical TCD scans.

Results of the parent BRIEF indicated that children in the abnormal TCD group exhibited greater executive dysfunction on the Inhibit and Shift scales in comparison to the normal and conditional groups. Additionally, the parents rated the conditional TCD group lower on Emotional Control than the normal TCD group. Even though the ratings on the parent BRIEF were statistically significant, the mean difference scores for the groups were not clinically significant (Kral & Brown, 2004). Results of the teacher BRIEF indicated that children in the abnormal group exhibited greater executive dysfunction on the Working Memory, Plan/Organize, and Monitor subscales. These results were statistically significant, but not clinically significant. The results of both the parent and teacher BASC rating scales did not yield statistically or clinically significant findings. The mean scores for the BASC parent and teacher ratings were within the average range (Kral & Brown, 2004).

Kral and Brown (2004) suggested additional research is needed to support parent and teacher ratings of behavior and children's neuropsychological functioning on the BASC and the BRIEF. They recommended this be done by using a clinically referred sample, because this type of sample would be expected to evidence heightened levels of executive functioning deficits, externalizing, or internalizing problems, or decreased adaptive functioning. Thus, the utility of

the BRIEF and BASC would be evidenced if such problems are noted in a clinically referred sample already exhibiting such problems.

The Current Study

Currently, there is limited research using the BRIEF and the BASC-2 to identify complications in children with SCD and no studies specifically including a clinically-referred sample of children and adolescents. So the primary aim of the current study will be to assess the criterion validity of the BRIEF and BASC-2 by using a clinically referred sample of children and adolescents with SCD. The clinically referred sample being examined was referred for behavioral and academic problems, which allowed for an evaluation of criterion validity of the BRIEF and BASC-2. It was predicted that the criterion validity would be supported for the BRIEF as evidenced by the participants from the clinically referred sample having elevated levels of executive dysfunction on the BRIEF's BRI and MI as compared to the normative sample of the BRIEF. It is also predicted that the criterion validity would be supported for the BASC-2 as evidenced by the participants from the clinically referred sample having elevated levels of behavioral and emotional problems and lowered adaptive functioning on the BASC-2's internalizing, externalizing, and adaptive domains as compared to the normative sample of the BASC-2.

A secondary aim will be to assess the criterion validity of the BRIEF and the BASC-2 based on stroke status of the clinically referred sample of children and adolescents with SCD. It is predicted that the criterion validity would be supported for the BRIEF as evidenced by the participants from the clinically referred sample with SCD and stroke evidencing more executive dysfunction as compared to those with SCD and without stroke. It is also predicted that the criterion validity would be supported for the BASC-2 as evidenced by the participants from the

clinically referred sample with SCD and stroke evidencing more behavioral problems as compared to those with SCD and without stroke.

CHAPTER 2: METHODS

Participants

This study used existing data from psychoeducational evaluations of 32 pediatric outpatients (21 males and 11 females) with various forms of SCD between the ages of 5 and 18 years that were seen between the months of March 2009 and June 2009. The children and adolescents were evaluated in a pediatric neuropsychology clinic for academic and attention problems. Inclusion criteria for the study included having SCD and no other chronic illness, being over 5 years of age and less than 18 years of age, being assessed at the clinic, and having completed a minimum of one parent or teacher BASC-2 or BRIEF.

Procedure

This research study was approved by East Carolina University's Institutional Review Board (see Appendix). Information from the teacher and caregiver BRIEF and BASC-2 forms were extracted from the clinic files. A demographic information sheet was created to extract basic participant information from the clinic files, which included age at evaluation, sickle cell type, sex, grade, and whether or not the child or adolescent was retained. Other information, such as the child's name, date of birth, address, or any other form of identification, was not extracted from the data.

To inspect the validity of the scores of the BRIEF and BASC-2, the validity indices of both measures were examined. Reports with validity indices in the caution and extreme caution ranges were flagged and further examined to determine if inclusion criteria were met for statistical analyses with the reports with validity indices in the acceptable range. Two raters examined all reports and each independently rated each report as valid or invalid based on the aforementioned criteria. The raters used available background information of the child, which

included medical records and teacher notes as well as other reports completed by teachers and parents of the child in question, to rate the possible validity of the report. The ratings of each report were compared and a final decision by two reviewers was made on which reports were considered valid for further statistical analyses.

Measures

BRIEF. The BRIEF is an 86 item questionnaire designed to assess difficulties in executive functioning in school-age children and adolescents using both parent and teacher forms (Gioia et al., 2000). It has 8 clinical scales which include Inhibit, Shift, Emotional Control, Initiate, Working Memory, Plan/Organize, Organization of Materials, and Monitor. These scales are then used to create the Behavioral Rating Index (BRI), the Metacognitive Index (MI), and an overall score, the Global Executive Composite (GEC). The BRI and MI were examined for the purposes of the current study. T-scores of 65 or higher are in the clinically significant range, 60-64 are in the at-risk range, and scores below 50 are in the normative range with a mean score of 50 for the measure. The T-scores of the BRIEF are normed for age and sex. The BRIEF has 2 validity indices: Negativity and Inconsistency. The Negativity Index measures the tendency to excessively rate the child as negative and the Inconsistency Index is based on similar items answered in an inconsistent manner (Gioia et al., 2000). Published test-retest reliability indicates that the composite scales are more reliable than the individual scales; thus, the current study focuses on the composite scales. Internal consistency reliability for the BRIEF is acceptable, ranging from .80 to .98 (Gioia et al., 2000). Toplak and colleagues (2009) examined the convergent validity of the BRIEF and performance-based measures of executive functioning, such as the Stop Task and Trail Making Task, in an ADHD sample and found evidence of high validity.

BASC-2. The BASC-2 is a questionnaire designed to assess behavioral and emotional problems, as well as adaptive functioning in children and adolescents (Reynolds & Kamphaus 2004). The BASC-2 has parent-, teacher-, and self-report forms, but only the parent and teacher forms (e.g., PRS and TRS respectively) were examined for the purposes of this study. The scales of the measures comprise the following composite scores: Externalizing Problems, Internalizing Problems, Behavioral Symptoms Index, and Adaptive Skills. For the externalizing and internalizing scales and Behavioral Symptoms Index, T-scores of 70 and above are in the clinically significant range, scores between 60 and 69 are in the at-risk range, and scores below 60 are in the normative range with a mean of 50 for the measure. For the adaptive scale, T-scores of 30 and below are in the clinically significant range, scores between 31 and 40 are in the at-risk range, and scores above 40 are in the normative range with a mean of 50 for the measure. The T-scores of the BASC-2 are normed for age and sex. Composite scales yielded higher test-retest reliability than the individual scales on both measures (PRS: .78 to .92 and TRS: .81 to .93). Only the composite scales were used in the current study. Correlations between the Conners Rating Scale-Revised and the TRS are moderate to high (Reynolds & Kamphaus, 2004). When comparing concurrent validity of the TRS to the TRF of the ASEBA, there were high correlations for externalizing problems (.75-.85) and internalizing problems (.64 to .80) between the two measures. When comparing concurrent validity of the PRS to the parent report form of the ASEBA, there were also high correlations for externalizing problems (.73. to .84) and internalizing problems (.65 to .75) between the two measures (Reynolds & Kamphaus, 2004).

The BASC-2 has 3 validity indices: F, Response Pattern, and Consistency. For the purposes of this study, the F and Consistency indices were examined. The F Index measures the

tendency to excessively rate the child as negative and the Consistency Index is based on response patterns to items that should be answered similarly (Reynolds & Kamphaus, 2004).

Data Analysis

Descriptive statistics for the parent and teacher ratings of the BRIEF and BASC-2 were reported. The validity indices of the BRIEF and BASC-2 were used to identify whether raters' reports appear overly negative or inconsistent. Identified reports were examined in further detail to determine if they were valid and could be used for the remaining analyses.

It was predicted that criterion validity would be supported for the BRIEF as evidenced by the participants from the clinically referred sample having elevated levels of executive dysfunction on the BRIEF's BRI and MI as compared to the normative sample. It was also predicted that criterion validity would be supported for the BASC-2 as evidenced by the participants from the clinically referred sample having elevated levels of internalizing and externalizing problems and lower adaptive functioning on the BASC-2 as compared to the normative sample. Mean scores and standard deviations were calculated for the two measures in regards to the primary aim. The primary aim was tested by using unpaired *t*-tests to compare the means of the T-scores of the BRIEF and BASC-2 to the criterion T-score of 50. The T-score of 50 was chosen to determine how far this population deviated from the normative mean. A chi-square analysis was used to examine the percent of ratings that fell into the at-risk and clinical ranges on the BRIEF and BASC-2 versus 16%, which is the expected percentage of the normative sample within this classification on one tail of the normal curve.

It was also predicted that criterion validity would be supported for the BRIEF as evidenced by the participants from the clinically referred sample with SCD and stroke evidencing more executive dysfunction as compared to those with SCD and without stroke.

Additionally, it was predicted that criterion validity would be supported for the BASC-2 as evidenced by the participants from the clinically referred sample with SCD and stroke evidencing more behavioral problems as compared to those with SCD and without stroke. The secondary aim was tested using unpaired *t*-tests to compare the means of the T-scores of the participants with SCD and stroke to the means of the T-scores of participants with SCD and without stroke. A chi-square analysis was then used to examine the percent of parent and teacher ratings in the at-risk or clinically significant ranges on the BRIEF and BASC-2 for the participants with SCD and stroke and the participants with SCD and without stroke.

CHAPTER 3: RESULTS

Descriptive Statistics of Participants

Of the 32 participants, 21 (65.6%) were male and 11 (34.4%) were female. The mean age of the participants was 9.6 years ($SD = 3.19$, range = 5.0 to 17.0 years) and the participants' grades ranged from kindergarten to twelfth grades. Twenty (64.5%) of the 32 participants had HbSS, seven (22.5%) had HbSC, four (12.9%) had HbS beta thalassemia-plus, and one participant's SCD type was not identified in the medical record. Twenty two (75.9%) of the 32 participants did not have a history of stroke, seven (24.1%) had a history of stroke, and for three participants, it could not be determined if they had a history of stroke. Approximately half of the children were previously retained a grade (48.3%; $n = 14$).

Validity of the Measures

Twenty one caregivers and 18 teachers completed the BRIEF. Sixteen (76.2%) of the 21 caregivers that completed the parent BRIEF were mothers, two (9.5%) were biological fathers, and 3 (14.3%) were foster mothers, grandmothers, or aunts. One hundred percent of the caregivers scored in the acceptable range on the Negativity Index. However, one caregiver (4.8%) scored in the caution range on the Inconsistency Index. After raters examined the individual parent BRIEF report flagged, it was deemed valid for analyses. One hundred percent of the teachers scored in the acceptable range on the Negativity and Inconsistency Indices. Thus, 21 parent BRIEFs and 18 teacher BRIEFs were retained for further analyses.

Twenty six caregivers and 20 teachers completed the BASC-2. Twenty one (80.1%) of the caregivers that completed the parent BASC-2 were mothers, two (7.7%) were biological fathers, and three (11.5%) were foster mothers, grandmothers, or aunts. One hundred percent of the caregivers scored in the acceptable range on the F Index. However, four caregivers (15.4%)

scored in the caution range and one caregiver (3.8%) scored in the extreme caution range on the Consistency Index. After examining the individual parent BASC-2 reports flagged, two of five reports were deemed valid. Two teachers (10%) scored in the extreme caution range on the Consistency Index. On the F index, three teachers (15%) were in the caution range and one teacher (5%) was in the extreme caution range. After examining the individual teacher BASC-2 reports flagged, four of six reports were deemed valid. Thus, 23 parent BASC-2s and 18 teacher BASC-2s were retained for further analyses.

Of the 30 participants used in remainder of the analysis, 21 (70.0%) were male and 9 (30.0%) were female. The mean age of the participants was 9.5 years ($SD = 3.27$, range = 5 to 17 years) and the participants grades ranged from kindergarten to twelfth grade. Eighteen (62.1%) of the 30 participants had HbSS, 7 (24.1%) had HbSC, 4 (13.8%) had HbS beta thalassemia-plus, and 1 participant's SCD type was not identified in the medical record. Twenty one (77.8%) of the 30 participants did not have a history of stroke, 6 (22.2%) had a history of stroke, and for 3 participants, it could not be determined if they had a history of stroke. Just under half of the children were previously retained a grade (48.2%; $n = 13$).

Comparison of BRIEF and BASC-2 Scores to Criterion Score of 50

It was predicted that the participants would have elevated levels of executive dysfunction on the BRIEF's BRI and MI as compared to the normative sample, which would support criterion validity of the BRIEF. It was also predicted that the participants from the clinically referred sample would have elevated levels of internalizing and externalizing problems and lower adaptive functioning on the BASC-2 as compared to the normative sample, which would support criterion validity of the BASC-2. The mean of the participants' T-scores on the BRI and MI of the BRIEF and externalizing, internalizing, and adaptive scales of the BASC-2 were significantly different than the T-score of 50 in every comparison (see Table 1). Specifically, the

participants' means were significantly higher than 50 on the BRI and MI of the parent and teacher BRIEFs, the externalizing and internalizing scales of the parent and teacher BASC-2s, and the school problems scale of the teacher BASC-2. Also, the participants' means were significantly lower than 50 on the adaptive scales of the parent and teacher BASC-2s.

Table 1. Summary of T-tests to Examine the Statistical Significance of BRIEF and BASC-2 Scores to Criterion T-Score of 50

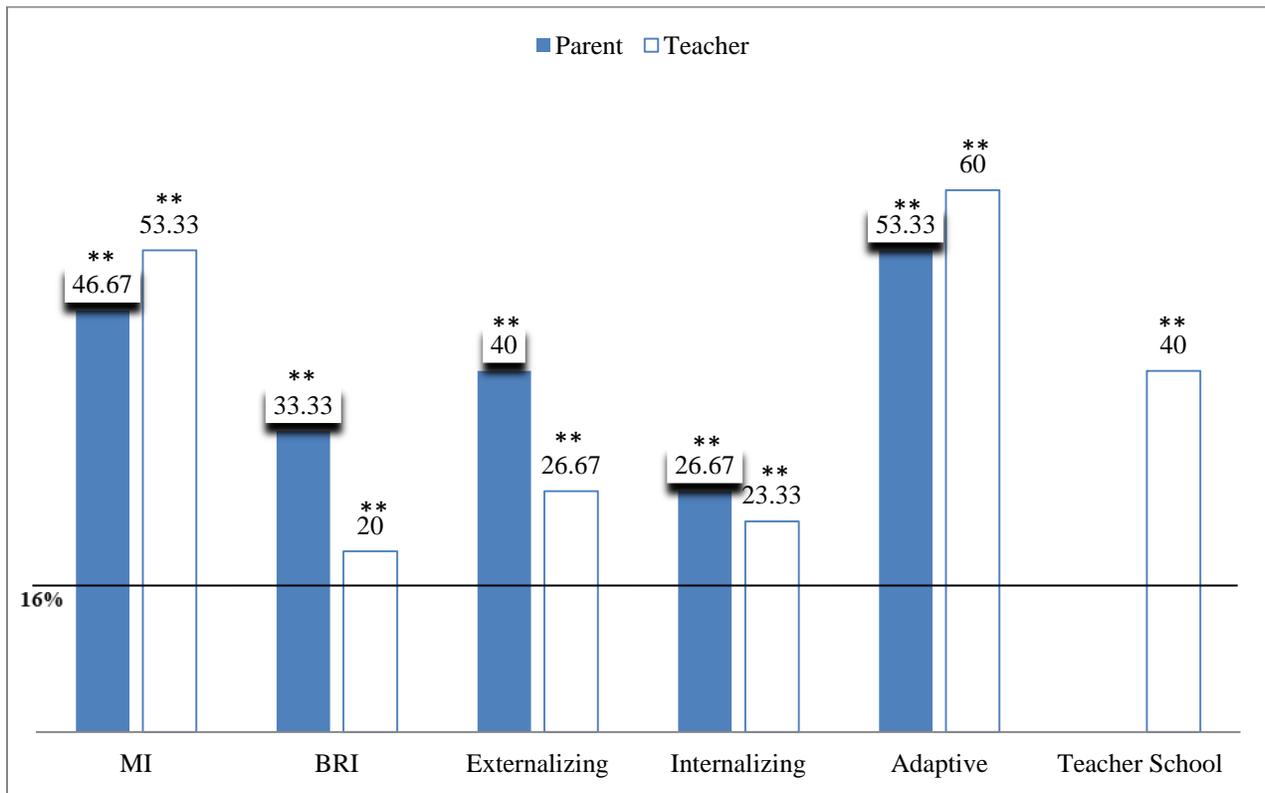
Measures	Mean	SD	Range
Parent BRIEF (<i>n</i> = 21)			
BRI	59.19*	11.37	42.00 – 77.00
MI	62.57**	11.34	42.00 – 84.00
Teacher BRIEF (<i>n</i> = 18)			
BRI	59.89**	14.23	42.00 – 89.00
MI	68.39**	9.04	47.00 – 83.00
Parent BASC-2 (<i>n</i> = 23)			
Externalizing Problems	56.85**	11.83	36.00 – 85.00
Internalizing Problems	56.27**	10.49	41.00 – 96.00
Adaptive Skills	43.38**	11.21	26.00 – 68.00
Teacher BASC-2 (<i>n</i> = 18)			
Externalizing Problems	56.80*	13.81	41.00 – 82.00
Internalizing Problems	57.15*	14.31	39.00 – 85.00
School Problems	62.63**	9.95	37.00 – 80.00
Adaptive Skills	40.65**	7.50	25.00 – 62.00

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

Note: BRI = Behavioral Regulation Index, MI = Metacognition Index

A chi-square analysis was then used to examine the percent of ratings that fell into the at-risk and clinical ranges on the BRIEF and BASC-2 versus 16%. The percentage of participants with at-risk and clinical ratings on the parent BRIEF ranged from 33.3% to 46.7%, on the teacher BRIEF ranged from 20% to 53.3%, on the parent BASC-2 ranged from 26.7% to 53.3%, and on the teacher BASC-2 ranged from 26.7% to 60%. When the percentage of participants' ratings in the at-risk and clinical ranges on the BRIEF and BASC-2 were compared to 16%, which is the standard percentage of the normative sample within these classifications, a higher percent of participants fell in the at-risk and clinical ranges in every comparison (see Figure 1). Of note, the highest percentage of ratings in the at-risk and clinical ranges on the parent and teacher BRIEF were on the MI scale and on the parent and teacher BASC-2 were on the Adaptive scale.

Figure 1. Percentage of At-Risk and Clinical Range Scores on the BRIEF and BASC-2 vs 16%



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

Comparison of Participants with and without Stroke

It was predicted that the participants from the clinically referred sample with SCD and stroke would evidence more executive dysfunction as compared to those with SCD and without stroke, which would support criterion validity of the BRIEF. It was also predicted that the participants from the clinically referred sample with SCD and stroke would evidence more behavioral problems as compared to those with SCD and without stroke, which would support criterion validity of the BASC-2. The mean of the participants with a history of stroke's T-scores on the BRI and MI of the BRIEF and externalizing, internalizing, and adaptive scales of the BASC-2 were not statistically different from each other (see Table 2). A chi-square analysis was then used to examine the percent of parent and teacher ratings in the at-risk or clinically significant ranges on the BRIEF and BASC-2 for the participants with SCD and stroke and the participants with SCD and without stroke. When the percentage of participants with SCD and with a history of stroke's ratings in the at-risk and clinical ranges on the BRIEF and BASC-2 were compared to the percentage of participants with SCD and without a history of stroke's ratings in the aforementioned ranges, they were not statistically different (see Figure 2). These analyses should be interpreted with caution due to the low number ($n = 6$) of participants with SCD and a known history of stroke.

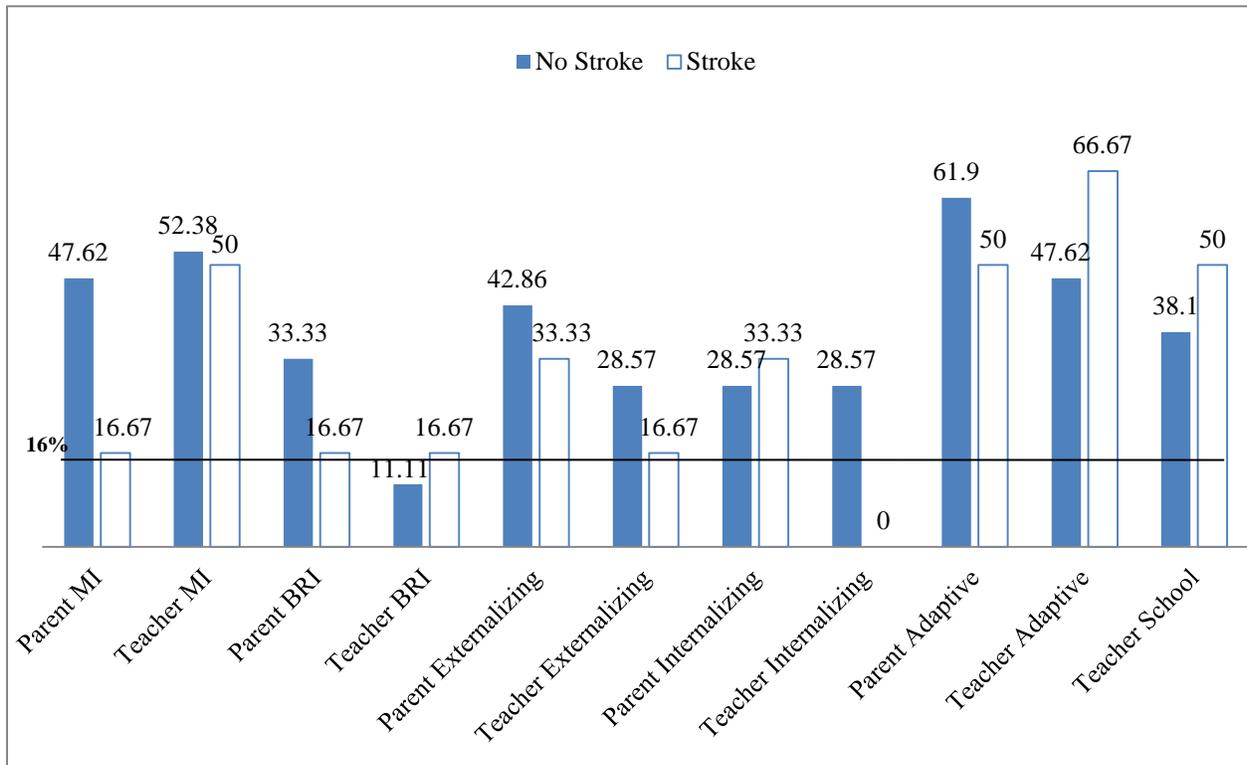
Table 2. Summary of Unpaired T-tests to Examine the Statistical Significance of the BRIEF and BASC-2 Scores of Participants of Different Stroke Statuses

Measures	No History of Stroke (<i>n</i> = 21)			History of Stroke (<i>n</i> = 6)		
	M	SD	Range	M	SD	Range
Parent BRIEF						
BRI	58.31	11.41	42.00 – 77.00	58.50	4.95	55.00 – 62.00
MI	60.88	11.85	42.00 – 84.00	60.50	3.54	58.00 – 63.00
Teacher BRIEF						
BRI	58.69	14.17	42.00 – 89.00	55.33	16.17	46.00 – 74.00
MI	68.77	10.41	47.00 – 83.00	65.67	4.51	61.00 – 70.00
Parent BASC-2						
Externalizing Problems	56.90	11.28	38.00 – 85.00	51.75	11.95	36.00 – 62.00
Internalizing Problems	56.65	11.03	46.00 – 96.00	54.00	11.69	41.00 – 67.00
Adaptive Skills	42.50	11.28	26.00 – 65.00	48.75	13.25	40.00 – 68.00
Teacher BASC-2						
Externalizing Problems	57.93	14.67	43.00 – 82.00	51.75	11.64	41.00 – 65.00
Internalizing Problems	58.50	16.60	39.00 – 85.00	52.25	7.04	42.00 – 58.00
School Problems	63.85	8.46	47.00 – 80.00	58.58	15.95	37.00 – 71.00
Adaptive Skills	40.00	5.96	25.00 – 49.00	45.25	11.87	34.00 – 62.00

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

Note: BRI = Behavioral Regulation Index, MI = Metacognition Index

Figure 2. Percentage of At-Risk and Clinical Range Scores on the BRIEF and BASC-2 of Children with SCD with and without Stroke



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

CHAPTER 4: DISCUSSION

The primary aim of the present study was to assess the criterion validity of the BRIEF and BASC-2 for children with SCD by using a sample of children with SCD referred for behavioral and academic problems. Results provide preliminary support that the parent and teacher BRIEF and BASC-2 are valid for identifying at-risk and clinically significant behavior and executive functioning problems in children with SCD. Specifically, participants from the clinically referred sample evidenced elevated levels of executive dysfunction on the parent and teacher BRIEF as compared to the normative sample of the BRIEF, and a higher percentage scored in the at-risk and clinically significant range than the normative sample. These findings are consistent with findings from Kral & Brown (2004) that indicated that the BRIEF is able to detect deficits in executive function among children with the HbSS genotype. Also, participants from the clinically referred sample had elevated levels of behavioral problems on the parent and teacher BASC-2 as compared to the normative sample of the BASC-2, and a higher percentage scored in the at-risk and clinically significant range than the normative sample. These findings support the BASC-2's use in detecting externalizing problems and adaptive problems in children with SCD.

Overall, these findings support the use of the BRIEF and the BASC-2 for assessing behaviors of concern for children with SCD in the school system. Psychologists working with children with SCD will have more tools to accurately identify problem areas, the severity of problems, and whether the children are in need of intervention. These measures may also be useful for the development and progress monitoring of targeted interventions. There are currently checklists that provide teacher perceptions of behavior, such as direct behavior ratings (DBR: Chafouleas, Riley-Tillman, & Sugai, 2007), but they only provide a limited scope of the

perceived level of behavior. The three target behaviors a DBR measures are disruptive behavior, academic engagement, and compliance/respect (Chafouleas, Riley-Tillman, & Sugai, 2007). Although DBRs may be used to identify children in need of behavioral interventions and assesses the severity of behaviors in the classroom, they do not identify the type or cause of problem areas. The BRIEF and BASC-2 may be able to better pinpoint the type and cause of behaviors noted by teacher ratings. After the BRIEF or BASC-2 is used to identify the problem area, DBRs may be used to gather baseline data and to document the effectiveness of the intervention daily, based on teacher report. Lastly, the BRIEF or BASC-2 can be given at the end of the intervention to obtain posttest data of the intervention's effectiveness.

The secondary aim of the study was to assess the criterion validity of the BRIEF and BASC-2 in detecting the differences in executive functioning and behavior problems in children with SCD with and without stroke. Findings indicated that neither the BRIEF nor the BASC-2 distinguished between children with SCD with and without stroke. Given research indicating more children with SCD and stroke experience executive dysfunction and behavior problems than children with SCD and without a history of stroke (Berkelhammer et al., 2007; Schatz et al., 2001), these findings provide preliminary evidence that the BRIEF and BASC-2 have limited validity for detecting the effects of stroke in children with SCD. Of note, the small sample size of participants with a known history of stroke may have contributed to these findings. Additionally, as medical records could not be obtained for some of our participants, it was probable that individuals who have a positive history of stroke were not included in the secondary aim analyses. This limitation may have attributed to the lack of significant findings to support the hypotheses involving participants with stroke and SCD.

Limitations

In addition to some of the limitations mentioned previously, other limitations are noted that should be addressed in future research. Although this study addressed the need to use the BRIEF and BASC-2 in a clinically referred sample as a way of evaluating validity for children with SCD, this study's limited sample size may have decreased the statistical power of the analyses. An additional replication with more participants should be a focus of future research to further validate the utility of the BRIEF and BASC-2 in children and adolescents with SCD. Statistical power was also decreased due to every child not having both forms (parent and teacher) of the BRIEF and BASC-2 completed. All measures were not obtained for every participant due to not completing measures keeping appointments, scheduling, and resistance from both parents and teachers. Future studies may need to provide an incentive so that teachers and parents are more likely to complete the forms when asked.

Future Directions and Clinical Implications

Future directions for research include continuing to validate the BRIEF and BASC-2 in children with SCD and other pediatric populations to determine if they are more sensitive than current measures for identifying executive functioning and behavior problems. Also, future studies on those already in existence should try to include a larger subset of participants with chronic illnesses when norming since these populations are also seen in the school system, and are more prone to school-related problems (Daly, Kral, & Brown, 2008). School psychologists need to stay abreast of new measures that can assess potential behavior problems because the new measures may be less expensive, more sensitive, or more time efficient than current measures commonly used in the schools. Clinicians may also want to try to validate the BRIEF in future studies because of the brevity of this measure compared to a full neuropsychological evaluation.

Lastly, school psychologists may want to conduct trials to determine if the BRIEF and the BASC-2 can be used as indicators of improvements for their interventions on children with chronic illnesses.

Future studies should administer either a TCD or MRI prior to administration of the BRIEF and BASC-2 to determine if they can detect stroke related differences in executive functioning and behavior. This is particularly important because children with SCD can have a stroke at any time. Using the BRIEF and BASC-2 in a larger sample of children with SCD with and without stroke will help to more accurately determine if the BRIEF or BASC-2 are useful tools for detecting stroke in this population. Also, since the study sample was referred for behavior and academic problems, a ceiling effect may have led to less variability between children with SCD with and without stroke. Thus, future research should use referred and non-referred children and adolescents.

School psychologists strive to be preventive through early detection of disabilities and academic deficits. This study was unable to determine the stroke status of participants based on BRIEF and BASC-2 scores, indicating there is a need to give every child with SCD a TCD or MRI at the beginning of each school year to determine if the child has experienced a stroke. Children with SCD are more likely to have evidence stroke, as a result early detection of stroke through TCD or MRI would be a possible way to identify students in need of treatment for stroke-related issues. Teachers and school psychologists should be notified if the TCD or MRI scans reveal a history of stroke so that they will be aware that the child may exhibit some academic or behavioral difficulties as a result. Then, if the children who have experienced a stroke begin to exhibit academic or behavior problems, an IEP or 504 meeting should be held to determine if special education services or certain accommodations may assist in the child's

educational progress. To provide data to support whether or not these services would be warranted, the Parent and Teacher BRIEF and BASC-2 should be administered to pinpoint the type and cause of problems behavior, if any. The school psychologist will be able to then use all of this information to help tailor an intervention to address the specific needs of that child with SCD and a prior history of stroke.

Clinical psychologists commonly use measures that are not often used in the schools. The more measures clinical and school psychologists both commonly use, the easier it will be to relay information or results back and forth between hospitals and academic settings that all parties understand. Collaboration between clinical psychologists and school psychologists is essential for the well-being of children with SCD because of the many medical complications associated with the disease. For example, since school psychologists do not administer full neuropsychological batteries, the BRIEF may be used as an alternative measure to get similar data. The child's clinician may view these BRIEF results and want to conduct other tests to further document the origin of the deficit or simply use the results of the BRIEF to help the parents come up with strategies that may be used in the home while the school psychologist comes up with strategies to be used in the school. School psychologists will be able to better consult and provide strategies with teachers, parents, and to the child with SCD if the results of the BRIEF and BASC-2 are used as tools to identify specific areas of concerns that can be addressed.

Conclusion

Overall, these findings support the BRIEF and BASC-2 as two measures that can be utilized with children with SCD as a means to examine executive functioning and behavior problems. The BRIEF and the BASC-2 provide school psychologists with measures equipped to

assess problems relating to multiple environments and day-to-day changes. The BRIEF and BASC-2 are also cheaper than a full neuropsychological battery, are quicker to administer, and provide more data addressing the type of deficits than other screening tools, such as DBRs. There are children with all types of illnesses in our educational system and it is our responsibility as their advocates to continue to conduct validity studies with every subgroup of children to help support the use of various assessment tools, such as the BRIEF and BASC-2, that accurately predict executive functioning and behavioral concerns in children so that we can provide the adequate support to further their academic progress. Through early identification, children can receive support through interventions at an earlier age. By being more proactive, rather than reactive, school psychologists will not be waiting for the child to fail, but assisting in the needs of children living with SCD.

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Appendix A: IRB Approval

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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: [Lee Purvis](#)
CC: [Cecelia Vairie](#)
Date: 7/13/2012
Re: [CR00000476](#)
[UMCIRB 11-0449](#)
[IMPORTED] Cognitive and Socioemotional Functioning 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 7/13/2012 to 7/12/2013. This research study is eligible for review under expedited category #5. 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
Protocol History	Study Protocol or Grant Application	7/6/2012 4:06 AM	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 IRB000004973