

Social Skills and Executive Function Among Youth With Sickle Cell Disease: A Preliminary Investigation

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Received June 19, 2013; revisions received December 2, 2013; accepted December 5, 2013

Objectives To explore the relationship between executive function (EF) and social skills in youth with sickle cell disease (SCD). **Methods** 20 youth with SCD completed objective tests of EF (Tasks of Executive Control; Animal Sorting subtest from the Developmental Neuropsychological Assessment-Second Edition), an IQ screener, and paper-and-pencil measures of social skills (Social Skills Improvement System [SSIS]). Primary caregivers completed paper-and-pencil measures of EF (Behavior Rating Inventory of Executive Function) and social skills (SSIS). **Results** EF scores from the Behavior Rating Inventory of Executive Function related to parent- and child-reported social skills such that EF deficits correlated with poorer overall and domain-specific social skills. Similarly, EF scores from the Animal Sorting test related to child-reported social skills. Worse parent-reported EF predicted worse parent-reported social skills above the variance accounted for by IQ. **Conclusions** EF is related to social skills and may be necessary for successful social interaction among youth with SCD. These results provide rationale and guidance for future larger-scale investigations of EF and social skills among children with SCD.

Key words executive function; sickle cell disease; social skills.

Sickle cell disease (SCD) is a genetically inherited chronic condition affecting 1 in 500 African American newborns in the United States (National Heart, Lung, and Blood Institute, 2009). Children and adolescents with SCD are at risk for social-emotional, neurocognitive, and academic functioning deficits (Hijmans et al., 2009; Lemanek & Ranalli, 2009; Unal, Toros, Kutuk, & Uyaniker, 2011), which may lead to the lower quality of life experienced by these youth (Palermo, Schwartz, Drotar, & McGowan, 2002). Youth with SCD have documented difficulties with social skills and peer relationships. Specifically, in studies assessing social relationships, females with SCD were less likely to be picked as a best friend, have reciprocated friendships, and were accepted less often than healthy

children (Noll et al., 1996; Noll, Reiter-Purtill, Vannatta, Gerhardt, & Short, 2007). Additionally, social competence in children and adolescents with SCD is significantly below average (Rodrigue, Streisand, Banko, Kedar, & Pitel, 1996). However, it is not clear that children with SCD always suffer social deficits. A longitudinal study by Noll, Kiska, Reiter-Purtill, Gerhardt & Vannatta (2010) assessing social functioning in children with SCD using peer ratings found that their social reputation was not impaired relative to their healthy peers.

Further research investigating more specific predictors of social functioning in youth with SCD is the next step toward clarifying these seemingly disparate findings. For example, social deficits in youth with SCD have been

investigated in the context of environmental factors, including limited access to peers and decreased school attendance, but additional etiologies of social skill deficits have not been fully explored (Bonner, Gustafson, Schumacher, & Thompson, 1999; Gustafson, Bonner, Hardy, & Thompson, 2006; Morgan & Jackson, 1986; Palermo, Schwartz, Drotar, & McGowan, 2002).

Given the relationship between social interaction and neurocognitive function (for a review, see Riggs, Jahromi, Razza, Dillworth-Bart, & Mueller, 2006), it has been hypothesized, but not tested, that social skills deficits in children with SCD may be related to difficulties with executive function (EF) including inattention and impulsivity (Gustafson, Bonner, Hardy, & Thompson, 2006). EF is involved in planning, initiation, purposive action, and effective performance (Lezak, 2004). Compared with healthy controls, youth with SCD without history of overt or silent stroke are more likely to show deficits in EF including concentration (Nabors & Freymuth, 2002), sustained attention (Brown et al., 1993; Noll et al., 2001), freedom from distractibility (Steen et al., 2003), planning, visual-spatial working memory (Hijmans et al., 2011), initiation, and organization (Berg, Edwards, & King, 2012). Youth with SCD are at a particular risk for these EF deficits compared with other neurocognitive deficits due to their chronic anemia and the associated autoregulation of cerebral blood flow (Venketasubramanian, Prohovnik, Hurlet, Mohr, & Piomelli, 1994). In addition, SCD affects the oxygen-carrying capacity of hemoglobin (Serjeant, 1992). The white-matter-rich frontal lobes are susceptible to this decreased oxygenation, and therefore, functions served by the frontal lobes (e.g., EF) are affected (Kral et al., 2003; Prohovnik et al., 1995). EF deficits are among the most common cognitive challenges among youth with SCD, even those without cerebral infarcts (Hijmans et al., 2011; Schatz & Roberts, 2007).

These deficits in EF may alter the way youth with SCD engage in or use social networks, thus leading to increased feelings of social isolation and distress (Bonner, Gustafson, Schumacher, & Thompson, 1999; Gustafson, Bonner, Hardy, & Thompson, 2006; Morgan & Jackson, 1986; Palermo, Schwartz, Drotar, & McGowan, 2002). Research in other pediatric populations suggests that there is an association between EF and social skills. Wolfe and colleagues (2012) found that child and adolescent brain tumor survivors with EF deficits also had poorer parent-reported and self-reported social skills. Additionally, children who have sustained a traumatic brain injury have poorer parent-reported social skills and difficulty generating strategies to solve social conflicts, a component of EF (Janusz, Kirkwood, Yeates, & Taylor, 2002). Further,

research in developmental literature suggests that children with attention-deficit/hyperactivity disorder with greater inattention and executive deficits also experience difficulties with peers (Huang-Pollock, Mikami, Pfiffner, & McBurnett, 2009; Pelham & Milich, 1984).

Taken together, the literature is clear that a disproportionate number of children with SCD have neurocognitive deficits, specifically in EF, as these are functions guided by the frontal lobes that are particularly susceptible to effects from the SCD process. Further, current literature implies that some youth with SCD have deficits in social skills, while others may not, and the reasons for such heterogeneity in functioning are unclear. Social skills are shown to be affected by EF in other pediatric populations, but this link has not been documented among youth with SCD. Therefore, the primary aim of this small pilot study was to examine the correlation between EF and specific social skill domains, to provide rationale and guidance for future studies in this area. Youth with SCD and no documented overt or silent stroke, or abnormal cerebral blood flow, were administered objective measures of EF. In addition, these youth and their primary caregiver completed subjective paper-and-pencil measures of EF and social skills. We hypothesized that overall social skills (both parent- and child-report) would be related to global measures of both objective (i.e., neuropsychological measures) and parent-report of EF.

Method

Participants

Twenty youth (12 girls, 8 boys) ranging in age from 8 to 16 years ($M = 11.25$, $SD = 2.8$) with SCD without known prior neurologic injury participated in the study. Participants were recruited during consecutive clinic visits during the recruitment period from a large tertiary care childhood medical center in the southeast United States. One family declined to participate. Participants' genotypes include the following: HbSS ($N = 14$); HbSC ($N = 4$); HbSB + Thal ($N = 1$); HbSB0 Thal ($N = 1$). Inclusion criteria included (1) diagnosis of SCD; (2) if magnetic resonance imaging screening was available ($N = 14$), no evidence of overt or silent infarct; and (3) between the ages of 8–16 years at the time of the study. Exclusion criteria included (1) $IQ \leq 70$ and (2) conditional or abnormal transcranial doppler screening. This study was approved by the university institutional review board (IRB). All parents or guardians provided IRB-approved informed consent. Participants also completed informed consent (ages 14–16 years) or assent forms (ages 8–13 years) per IRB protocol. Informants for the caregiver questionnaires were mostly mothers (95%),

and one aunt (5%), though there was no formal exclusion of male guardians. All of the caregivers and their children were African American.

Measures

Social Skills

Child social skills were measured with the Social Skills Improvement System (SSIS; Gresham & Elliot, 2008), a measure assessing skill domains necessary for successful social interaction. This measure includes the domains of Communication, Cooperation, Assertion, Responsibility, Empathy, Engagement, and Self-Control. Caregivers were asked to rate how true the behavior is for their child (e.g., Overreacts to small problems; Needs to be told to begin a task even when willing; Makes careless errors). Each question is rated on a 4-point scale ranging from "not true" to "a lot true." Responses are summed and standardized by age for a Social Skills Summary score. Higher scores indicate better social skills. This measure has excellent internal consistency reliability (Cronbach's $\alpha = .95$ for overall scale), as well as modest to high correlations with other social skills measures (e.g., Social Skills Rating System, Behavior Assessment System for Children, Second Edition; Gresham & Elliot, 2008). Each caregiver and child completed the SSIS rating scale.

Executive Function

EF was measured with several instruments. First, caregivers completed the Behavior Rating Inventory of Executive Function (BRIEF; Gioia, Isquith, Guy, & Kenworthy, 2000), an 86-item caregiver-report measure of everyday behavior in eight domains of EF. Three summary measures are calculated: Behavioral Regulation Index (BRI); Metacognition Index (MI); and Global Executive Composite (GEC). The BRI is a measure of ability to use inhibitory control to change behaviors and emotions. The MI is a measure of ability to monitor tasks and performance. The GEC is a summary measure of all subscales. Scores are converted to T-scores with higher T-scores indicating greater dysfunction. This measure has high internal consistency for both the full scale (GEC Cronbach's $\alpha = .97$) and subscales (Cronbach's $\alpha \geq .80$). It also has high construct validity as measured by correlation with general behavioral functioning measures.

Second, children and adolescents ($N = 14$) completed the Tasks of Executive Control (TEC; Isquith, Roth, & Gioia, 2010), a computerized measure assessing inhibitory control, working memory, vigilance, and sustained attention. Participants completed six levels including zero-back with and without inhibition of stimuli; one-back with and without inhibition; and two-back with and without

inhibition. Results are converted to age-normed T-scores with higher scores indicating greater impairment. Scores across all levels provide summary scores in several domains including Sustained Accuracy (accuracy in responding), Response Speed, and Response Speed Intraindividual Coefficient of Variability (ICV; consistency of response speed). Internal consistency for the TEC is high, with split-half reliabilities of $\geq .88$, $.98$, and $.71$ for Sustained Accuracy, Response Speed, and Response Speed ICV, respectively. Test-retest reliability is also high, ranging from $.71$ to $.80$ for the three domains. Further, the TEC domains correlate modestly with relevant subscales on the BRIEF (Isquith, Roth, & Gioia, 2010).

Third, children and adolescents completed one subtest from the Developmental Neuropsychological Assessment-Second Edition (NEPSY-II; Korkman, Kirk, & Kemp, 2007). The NEPSY-II is a standardized measure of neuropsychological functioning assessing several domains. Children and adolescents completed the Animal Sorting subtest within the attention/executive functioning domain, which required them to sort eight cards into two groups of four cards and measured their ability to sort into categories, plan, and shift set between concepts. Animal Sorting shows test-retest correlations of $.64$ – $.73$ depending on age, as well as shows higher correlations with subtests in the same domain (i.e., attention/executive functioning) than with subtests from other domains. Further, Animal Sorting is modestly correlated with other EF measures including the Delis-Kaplan Executive Function System (Korkman, Kirk, & Kemp, 2007).

Intelligence

Each participant's IQ was measured using the Wechsler Abbreviated Scale of Intelligence (WASI; Wechsler, 1999), a well-used intelligence measure, which has excellent reliability and validity in child populations. Two subtests were used as a screener for each participant's IQ: Vocabulary and Matrix Reasoning.

Medical Chart Review

A review of each participant's medical record was conducted to determine patient age, gender, ethnicity, and disease subtype.

Procedure

Assent and consent were obtained from the youth and the youth's caregiver. Following the consent process, each participant's caregiver completed a demographic questionnaire, the SSIS and the BRIEF. Children and adolescents with SCD completed Animal Sorting, the TEC, the

WASI, and the SSIS. Quantitative data were analyzed in SPSS Version 20 (IBM Corporation, 2011).

Results

Relations Between Executive Function and Social Skills

Given this study's small sample size and number of comparisons, we set alpha at .01 to control for Type I error. Data from the TEC (Sustained Accuracy, Response Speed, and Response Time ICV), SSIS Overall Social Skills, Animal Sorting, IQ, and BRIEF (GEC, BRI, and MI scales) are presented in Table I. Mean scores on each variable were within the average range with the exception of Animal Sorting, which was below average. To address the hypothesis that better scores on EF measures would correlate with higher scores on measures of social skills, we examined correlations between both child- and parent-reported SSIS Social Skills Summary scores and measures of EF (Table II). Two summary measures from the BRIEF (BRI and GEC) and Animal Sorting were related to child-reported social skills such that EF deficits were related to poorer social skills. Summary measures from the BRIEF (BRI, MI, and GEC) were all related to parent-reported overall social skills such that EF deficits correlated with poorer social skills ($r = -.81$, $-.90$, and $-.92$, respectively).

TEC variables and BRIEF MI were not significantly related to child-reported social skills. TEC variables and Animal Sorting were not significantly related to parent-reported social skills. Therefore, the BRIEF GEC and Animal Sorting were retained for further exploratory analyses

investigating relationships among EF and specific child-reported social skills; similarly, BRIEF GEC was retained for further analyses for parent-reported social skills.

BRIEF GEC was related to parent-reported social skills domains of Communication, Cooperation, Responsibility, Empathy, Engagement, and Self-Control (all r 's $\leq -.75$), such that worse EF was related to poorer specific social skills. In contrast, BRIEF GEC was only significantly related to the child-reported social skill domain of Self-Control ($r = -.67$), in the predicted direction. Animal Sorting was significantly related to the child-reported domains of Responsibility ($r = .60$) and Self-Control ($r = .64$), such that worse EF was related to poorer specific social skills.

Table II. Correlations Between Overall Social Skills and Executive Function Variables

| Measures | SSIS parent-report social skills | SSIS child-report social skills |
|-------------------------------|----------------------------------|---------------------------------|
| BRIEF BRI | -.81*** | -.69*** |
| BRIEF MI | -.90*** | -.52 |
| BRIEF GEC | -.92*** | -.62** |
| TEC Sustained Accuracy | -.61 | -.36 |
| TEC Response Speed | -.47 | .09 |
| TEC Response Time ICV | -.33 | -.48 |
| NEPSY-II Animal Sorting Total | .39 | .65** |

Note. BRIEF = Behavior Rating Inventory of Executive Function; BRI = Behavioral Regulation Index; MI = Metacognition Index; GEC = Global Executive Composite; TEC = Tasks of Executive Control; ICV = Intraindividual Coefficient of Variability; NEPSY-II = Developmental Neuropsychological Assessment-Second Edition; SSIS = Social Skills Information System.

** $p < .01$, *** $p < .001$.

Table I. Descriptive Data

| Measure | N | Mean (SD) | No. of impaired ^a (%) |
|---|----|--------------|----------------------------------|
| BRIEF BRI ^b | 20 | 54.3 (14.4) | 5 (25%) |
| BRIEF MI ^b | 20 | 55.3 (13.3) | 5 (25%) |
| BRIEF GEC ^b | 20 | 55.2 (13.9) | 4 (20%) |
| TEC Sustained Accuracy ^b | 14 | 60.9 (19.1) | 6 (43%) |
| TEC Response Speed ^b | 14 | 54.4 (16.1) | 3 (21%) |
| TEC Response Time ICV ^b | 14 | 59.5 (19.7) | 5 (36%) |
| NEPSY-II Animal Sorting Total ^c | 20 | 5.8 (2.9) | 9 (45%) |
| SSIS Parent-Report Social Skills ^d | 20 | 95.9 (20.3) | 4 (20%) |
| SSIS Child-Report Social Skills ^d | 20 | 106.2 (14.9) | 2 (10%) |
| IQ ^d | 20 | 89.7 (13.9) | 4 (20%) |

Note. BRIEF = Behavior Rating Inventory of Executive Function; BRI = Behavioral Regulation Index; MI = Metacognition Index; GEC = Global Executive Composite; TEC = Tasks of Executive Control; ICV = Intraindividual Coefficient of Variability; NEPSY-II = Developmental Neuropsychological Assessment-Second Edition; SSIS = Social Skills Information System; IQ = Intelligence Quotient.

^aImpairment defined as ≥ 1.5 standard deviations below the mean.

^bT-score ($M = 50$, $SD = 10$)

^cScaled score ($M = 10$; $SD = 3$)

^dStandard score ($M = 100$; $SD = 15$)

Prediction of Social Skills by IQ and Executive Function

Finally, we investigated whether social skills deficits could be explained by global IQ separately from EF difficulties. A series of hierarchical multiple regression analyses predicting both parent- and child-reported overall social skills were conducted. For parent-reported social skills, we entered IQ in Step 1 and BRIEF GEC in Step 2. This analysis indicated that worse parent-reported EF as measured by the BRIEF GEC predicted worse parent-reported social skills over and above the variance accounted for by IQ ($\Delta R^2 = .57, p < .001$). IQ was not a significant predictor of parent-reported social skills after accounting for a summary measure of EF. A second regression predicting child-reported social skills from IQ in Step 1 and BRIEF GEC and Animal Sorting in Step 2 was significant ($R^2 = .65, p = .001$), indicating that worse EF as measured by the BRIEF GEC and Animal Sorting predicted worse child-reported social skills over and above the variance accounted for by IQ.

Discussion

The present pilot study examined relationships between EF and social skills in children and adolescents with SCD. Both EF and social skills are multifaceted constructs and have been found to be related among other brain-injured populations (e.g., Janusz, Kirkwood, Yeates, & Taylor, 2002; Wolfe et al., 2012). In the present study, we identified parent-report of EF to be correlated with both overall and domain-specific parent-reported social skills. Similarly, parent-report of EF was also correlated with overall child-reported social skills, specifically for responsibility and self-control domains. The finding that EF is related to several social skills domains across informants and instruments suggests that deficits in EF could potentially have global implications for the development and maintenance of social functioning. Certainly aspects of EF, such as being able to sustain attention, switch behavior to meet changes in task demands, hold information in memory while formulating responses and processing social information quickly and accurately are necessary skills for successful social interaction, along with other characteristics including emotionality, sociability, and parental factors (Sanson, Hemphill, & Smart, 2004).

It is noteworthy that in the exploratory analyses, EF predicted parent-reported social skills above and beyond the contribution of general intellectual ability, suggesting the increased importance of specific variables related to social skills such as EF versus more global cognitive

abilities, a result that has been documented in other pediatric populations (Wolfe et al., 2012) and should be followed up in future studies with larger sample sizes. This study identified a discrepancy between subjective (i.e., parent-report) and objective (i.e., patient-completed tasks) measures of EF. Parent-report of EF was strongly related to parent-report of social skills, yet objective measures of EF were weakly or unrelated to parent-reported social skills. In turn, one objective measure of EF (Animal Sorting) was significantly related to child-reported overall social skills, and more specifically, to child-reported responsibility and self-control. The Animal Sorting subtest assesses cognitive flexibility and planning, and one can intuit how these abilities might relate to skills such as responsibility and self-control.

Although it is not surprising that parent-reported social skills and parent-reported EF were related given that they were completed by the same judge, these results are consistent with observations suggesting that parent-report and neuropsychological measures may only be modestly correlated (Anderson, Anderson, Northam, Jacobs & Mikiewicz, 2002; Mangeot, Armstrong, Colvin, Yeates, & Taylor, 2002). However, as mentioned above, youth self-report of social skills was related to both an objective and parent report measure of EF, indicating that perhaps school-age youth with SCD may have somewhat better insight than their parents into their strengths and weaknesses with regard to social skills.

The sample size in this study was notably small, and therefore results should be considered tentative, pending further investigation into the relationship between EF and social skills in this population. Future research should use larger samples of children with SCD to examine if the findings hold true. Additionally, the cross-sectional design in our study precludes causal direction between EF and social skills, although prior studies suggest that EF is a necessary precursor to social skill development (Carlson, Mandell, & Williams, 2004). Longitudinal research examining how these two constructs change over time as children mature will be necessary to determine causal relationships. Our study did not incorporate control-group comparisons, and thus we cannot make conclusions about the social functioning of children with SCD as compared with healthy peers. However, our sample's normed performance on measures of EF and social skills was generally within the average range, suggesting that although some children may experience deficits in these areas, many children do not. This is consistent with prior research using peer ratings to examine social functioning in youth with SCD (Noll, Kiska, Reiter-Purtill, Gerhardt & Vannatta, 2010; Rodrigue, Streisand, Banko, Kedar, & Pitel, 1996). Future research

would benefit from comparisons with control groups to determine functioning relative to healthy children, and controlling for variables such as socioeconomic status.

Given the nature of this pilot study, we were unable to obtain teacher report on children's social skill development. Given that teachers may have differing views of their student's peer relationships than parents, future research should incorporate teacher ratings of EF and social functioning. Further, our sample was composed of youth with SCD without overt stroke or abnormal cerebral blood flow. Approximately 30% of children with SCD will have cerebrovascular disease (DeBaun et al., 2012), and these children experience greater cognitive deficits than children without overt or silent stroke (Schatz, Brown, Pascual, Hsu, & DeBaun, 2001). Additional studies should investigate these relationships among more severely affected children with SCD. Finally, it should be noted that child-report of social skills and parent-report of EF may not be completely independent, thereby inflating the relationship between the measures due to rater bias. Future research should certainly make use of different informants and objective measures of EF and social skills.

Discernment of underlying neurocognitive processes necessary for successful social interaction is crucial to the development of interventions. Our study demonstrates that deficits in EF may partially explain the social difficulties evidenced by a subset of children and adolescents with SCD, which future research should further investigate. Thus, in addition to teaching specific social skills, targeting underlying EF, such as attention, working memory, and impulsivity may improve the efficacy of social skills interventions and lead to more sustainable and generalizable intervention outcomes for those youth with SCD who struggle with social functioning. Comprehensive neuropsychological evaluations for children with SCD, including in-depth assessment of EF, may provide valuable information to clinicians in identifying children who may also be at risk for social skills difficulties.

Funding

This research was funded in part by a training grant from the National Cancer Institute (5R25CA047888-22). Ms. Hensler receives financial support from this grant. No other authors have any financial disclosures.

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