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Responsive Parenting Behaviors and Cognitive Function in Children With Sickle Cell Disease

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Abstract

Objective Children with sickle cell disease (SCD) are at increased risk for cognitive impairment as a result in part from biological characteristics of the disease; however, limited research has explored possible social and contextual factors associated with risk for cognitive problems. The primary aim of the present study was to examine the relation between children's cognitive functioning and responsive parenting, a potentially important contextual factor in children with SCD, accounting for family socioeconomic disadvantage, child disease severity, and caregivers' perceived stress. **Methods** Forty-eight children completed standardized cognitive assessments and caregivers provided self-reports of general and disease-related stress. Parent–child dyads completed a video recorded puzzle-solving task and observed parenting was quantified using two coding systems. Bivariate Pearson correlations were used to assess preliminary hypotheses, and linear multiple regression analyses were used to assess the primary hypothesis. **Results** Results suggested that increased levels of parental stress were related to fewer observations of responsive parenting and provided evidence of an association between children's cognitive function and responsive parenting. Specifically, increased disease-related parent stress and reduced parental use of expansive language were associated with significantly lower cognitive functioning in children with SCD. **Conclusions** Findings suggest that social environmental factors along with disease characteristics are sources of risk for cognitive problems with children with SCD. Further, these findings highlight the need to develop targeted interventions for parents of children with SCD to decrease levels of stress and enhance parenting skills, with the aim improving cognitive functioning in youth.

Key words: cognitive function; parenting; sickle cell disease.

Sickle cell disease (SCD) is an inherited blood disorder that affects more than one in every 400 African-American in the United States (Hassell, 2010). Impaired cognitive function is one of the most significant negative developmental outcomes faced by individuals with SCD. Significant deficits have been found across several domains of cognitive function, including overall intellectual function, executive function, and academic achievement when compared to healthy peers and normative samples (e.g., Compas, Jaser, Reeslund, Patel, & Yarboi, 2017; Prussien, Jordan, DeBaun, & Compas, 2019). In spite of the cognitive

difficulties in children with SCD, factors contributing to increased risk for problems are not well understood. The current study examined responsive parenting behaviors in addition to other social-environmental factors (including family socioeconomic disadvantage and parent stress) that may play important roles in cognitive function in children and adolescents with SCD.

Most research exploring factors associated with cognitive impairment in children with SCD has focused on disease-related factors, particularly those that are associated with cerebral infarcts and insufficient cerebral oxygenation (Prussien et al., 2019). Many of the complications associated with SCD including observable and silent stroke, elevated cerebral blood flow, sleep-disordered breathing, and anemia are related to poorer performance on a range of cognitive tasks in affected children (e.g., Sanchez, Schatz, & Roberts, 2010). However, the risk for cognitive problems associated with disease characteristics needs to be considered along with characteristics of children's social environments (Brown, Doepke, & Kaslow, 1993).

First, families of children with SCD, who are predominantly African American, are faced with high levels of socioeconomic disadvantage and stress (Thompson, Gil, Burbach, Keith, & Kinney, 1993). U.S. Census Bureau data (DeNavas-Walt & Proctor, 2015) indicate that while the national poverty rate is 15%, African-Americans are disproportionately represented with an estimated 26% of families living below the poverty line. Consequently, many children with SCD and their families face poverty and chronic financial hardship (Panepinto, Pajewski, Foerster, Sabnis, & Hoffmann, 2009). For example, King et al. (2014) found that over 50% of individuals with SCD in the U.S. lived at or below poverty, and nearly 70% received Medicaid coverage. Yarboi et al. (2017) found maternal financial strain to be associated with lower self-reported positive parenting and several domains of cognitive function in children with SCD; and Schatz, Finke, and Roberts (2004) found that the effect of chronic anemia on cognition in children with SCD was dependent on their family's socioeconomic status, suggesting interdependence of these risk factors.

Second, both disease severity and family socioeconomic disadvantage may be associated with disruptions in parenting, in part through increased levels of actual and perceived parental stress. Caregivers in economically disadvantaged families are often faced with the challenges of unemployment; lack of vital resources including food, water, and heat; and low levels of family and community support (Middlemiss, 2003). Persistent exposure to financial hardship and limited social support are associated with chronic stress and distress in parents, including elevated symptoms of depression (e.g., Conners-Borrow et al., 2014). Further, the severity of a child's illness is related to increased levels of stress for parents. Brown et al. (2008) proposed that parents of chronically ill children, including those with SCD, are exposed to numerous stressors including financial strain, disruptions in daily routines, and high levels of caretaking responsibilities. And in a review, Cousino and Hazen (2013) found that caregivers of children experiencing pediatric illness (e.g., asthma, cancer, and diabetes) reported greater parenting stress than caregivers of healthy children.

In the context of SCD, research has shown that there is a significant inverse relation between disease-related parenting stress and family functioning among families with children with SCD, particularly with regard to parent–child communication about sickle cell-related issues (Barakat, Patterson, Tarazi, & Ely, 2007). Family income is also a significant predictor of disease-related parenting stress (Barakat et al., 2007). Importantly, illness parameters such as disease severity and duration may contribute more to parenting stress among caregivers of children with SCD than in other illness populations (Barakat et al., 2007; Logan, Radcliffe, & Smith-Whitley, 2002). Further, Bills, Schatz, Hardy, and Reinman (2019) found that parent and family functioning predicted language scores in children with SCD even after controlling for socioeconomic status. These studies suggest that parents of children with SCD, many of whom also face socioeconomic disadvantage, may experience a double hit to their capacity to provide warm, structured, and responsive parenting, due to additional disease-related stress.

The stress and negative emotions associated with economic disadvantage and other sources of stress compromise parents' abilities to engage in responsive interactions with their children (Bradley & Corwyn, 2002). Research has shown that economically disadvantaged caregivers engage in parenting behaviors that lack consistency and sensitivity to children's needs (e.g., Hackman, Farah, & Meaney, 2010; Luby et al., 2013), which manifests as disruptions in parent–child interactions (e.g., Evans et al., 2010). Research with healthy, typically developing children suggests that responsive parenting is a key factor in cognitive development in children (e.g., Doan & Evans, 2011). As such, the resilience displayed by some children raised in chronically disadvantaged environments may be attributed to parenting that is responsive and attuned to the children's needs. Responsive parenting includes the provision of consistently high levels of warmth and acceptance of their children, responses that are contingently linked to children's behavior, rich language input, and maintaining the children's interests may be especially

important for children's cognitive growth (Landry et al., 2012). These types of parenting behaviors may also protect children from other factors that impede cognitive development, including the effects of poverty. Therefore, it is important to consider the possible role of responsive parenting as a resource for cognitive development in children with SCD.

The current multimethod study used direct observations of parenting behaviors, parent reports of stress related to socioeconomic disadvantage and medical care for their children, and direct measures of children's cognitive function. Based on previous research, we examined measures of overall intelligence (IQ), executive function, and reading achievement as indicators of cognitive function (see Prussien et al., 2019, for review). The following preliminary hypotheses were tested: (a) Greater SCD severity and socioeconomic disadvantage will be associated with higher levels of parent stress; (b) higher levels of parent stress and socioeconomic disadvantage will be associated with lower levels of responsive parenting behaviors; (c) higher parenting stress, greater socioeconomic disadvantage, and lower levels of responsive parenting behaviors will be associated with lower scores on measures of cognitive function in children with SCD. Finally, the primary hypothesis of this study was that (d) parent stress and responsive parenting behaviors will be associated with children's cognitive function, above and beyond the variance accounted for by disease severity and socioeconomic disadvantage alone.

Method

Participants

Forty-eight children and adolescents with SCD ages 6 to 16 ($M = 9.30$; $SD = 3.08$ years) and their caregivers were enrolled in the study. Participants were 56.8% male and included four SCD subtypes: 70.8% were diagnosed with HbSS, 18.8% with HbSC, 6.3% with HbSβ^0 thalassemia, and 4.2% with HbSβ^+ thalassemia. The majority of children ($n = 38$; 77.3%) displayed no evidence of an overt cerebral infarct. Seven patients presented with a history of silent stroke and three with a history of overt stroke, and 52.1% of the sample had a history of acute chest syndrome. Sixty seven percent of the sample had an active prescription for hydroxyurea and 14.6% were on a chronic transfusion plan. Average hemoglobin level near the time of the study visit was 9.59 ($SD = 1.54$), and 45.8% of the sample had an emergency department visit within 6 months prior to the study visit. Seven percent repeated a grade, and 22.7% received special services at school (e.g., Individualized Education Program, 504 plan). The majority of the sample (97.7%) identified as African American.

Participants also included 48 primary caregivers of youth with SCD. Caregivers were primarily biological parents ($n = 42$), and included adoptive parents ($n = 1$), grandparents ($n = 3$), and other primary caregivers ($n = 2$). Caregivers reported spending daily face-to-face time with their child; thus, the terms "parent" and "caregiver" are used interchangeably throughout. Overall, caregivers ranged in age from 25 to 60 years ($M = 39.57$, $SD = 9.43$), and 79.5% were female. Caregivers came from a range of educational backgrounds (11th grade to 3rd year of graduate school; $M = 13.81$ years of education), as well as a range of annual family income levels (37.2% earned $25,000 or less, 30% earned $25,001 to $50,000, 16.3% earned $50,001 to $75,000, 9.3% earned $75,001 to 100,000, and 7.0% earned $100,000 or above).

Procedure

Families were recruited to participate in a study of cognitive function in children with SCD. Eligibility requirements included: (a) confirmed diagnosis of SCD, (b) child age of 6–16 years, and (c) participation of a caregiver who had legal guardianship of the child. Children with a history of comorbid neurologic disorder (e.g., neurofibromatosis and tuberous sclerosis) were excluded from participating; only one patient was excluded for this purpose.

Informed consent was obtained from caregivers and informed assent was obtained from children prior to study entry and participation. The study protocol was approved by the university's Institutional Review Board. Recruitment occurred at a university-based children's hospital and an affiliated community clinic in the southeastern United States where participants received their care. Eligible families were identified by members of the pediatric hematology medical team. After receiving verbal consent to be approached by a member of the research team, families were given additional information and were recruited for participation if interested. Eighty families expressed interest, and 48 participated. During the 2.5-hr laboratory-based session, children completed a 2-hr cognitive assessment while caregivers completed a series of questionnaires on family demographics and self-reported social-environmental stress and parenting stress. Caregiver-child dyads completed a 10-min video recorded interaction task that served as the source of parenting behaviors for coding.

Measures

Full Scale IQ

Children completed the Wechsler Abbreviated Scale of IQ, Second Edition (WASI-II; Wechsler & Hsiao-pin, 2011). The WASI-II is a valid and reliable measure widely used to assess IQ in children. The measure consists of four subtests: Block Design, Vocabulary, Matrix Reasoning, and Similarities; which are used to generate the Full-Scale IQ Quotient (FSIQ), a broad estimate of general intellectual ability.

Working Memory

Children were administered two subtests that make up the Working Memory Index (WMI) from the Wechsler IQ Scale for Children, Fourth Edition (WISC-IV; Wechsler, 2003). The WISC-IV is a valid and reliable measure and the benchmark test used to assess IQ in children. The WMI (Digit Span and Letter-Number Sequencing subtests) is a measure of the ability to concentrate, sustain attention, and exert mental control.

Reading

Children completed the Wide Range Achievement Test, Fourth Edition (WRAT4; Wilkinson & Robertson, 2006). The WRAT4 is a valid and reliable measure of the fundamental academic skills of reading, spelling, and math in children and adults. The current study focused on the Reading Composite score, which is a combination of examinees' performance on the Word Reading and Sentence Comprehension subtests.

Observed Parenting

Video-recorded interactions between caregivers and children were quantified using macrolevel and microlevel coding of parenting behavior and communication. As part of the 10-min task, dyads were asked to work together to complete a series of 15 tangram puzzles of increasing difficulty. Specifically, caregivers and children were instructed to "talk to each other and ask each other questions in order to solve the puzzles" as a measure of "how you and your child communicate and solve problems together." This direct observation of parenting is derived from similar paradigms used in previous studies of parenting and cognitive function (e.g., Deater-Deckard, Sewell, Petrill, & Thompson, 2010; Dunn et al., 2011; Landry et al., 2012). Video recordings of parent–child interactions were coded by trained graduate and undergraduate students. All coders were required to pass a written test of code definitions and examples and to train to 80% reliability on a series of standard recordings that have been previously recorded by expert raters. All observations were double-coded independently by two coders.

(1) Macrolevel. The Iowa Family Interaction Rating Scales (IFIRS) is a macrolevel coding system used to code caregivers' verbal and non-verbal communication, behaviors, and emotions in a videotaped interaction (Melby & Conger, 2001). Codes are assigned values from 1 to 9, with 1 reflecting the absence of the behavior or emotion and 9 indicating a behavior emotion is "mainly characteristic" of the caregiver during the interaction. The higher of two discrepant ratings was used when coders' ratings differed by one point. When ratings differed by two or more points, coders reached agreement through discussion. The Responsive Parenting composite was derived by summing scores from the following codes: stimulates cognitive development, encourages independence, listener responsiveness, communication, and sensitive/child-centered (see Table I for code descriptions and descriptive statistics). The Responsive Parenting composite was adapted based on a similar aggregation of IFIRS codes used in previous research to assess responsive parenting behaviors (Watson et al., 2014). Internal consistency of the codes that comprised the responsive parenting composite was $\alpha = 0.87$.

(2) Microlevel. The Contingency Coding System is a microlevel coding system used to code caregivers' conversational discourse with their children (Rodriguez et al., 2013). Caregivers and children's speech during the interaction task were transcribed into utterances. An utterance is defined as "a unit of speech with complete semantic and syntactic content" (McLaughlin, Schutz, & White, 1980). The first two parent utterances following each child utterance were coded and percentages of each code were calculated out of the total number of coded utterances. Any discrepancy among raters in individually assigned codes was discussed until coders reached agreement on one code. The current analyses include utterances coded as either topic maintenance or expansions (see Table I for code descriptions).

Socioeconomic and Disease Risk

Socioeconomic and disease risk factors were examined with two cumulative risk (CR) variables. Parents provided demographic information, including age, education level, race, annual family income, and marital status. Following an approach described by Bemis et al. (2015), each variable was dichotomized such that participants received a score of 0 or 1, indicating lesser or greater risk: caregiver partnered (0) versus single (1); annual family income $>$ \$25, 000 (0) versus $\leq$ \$25, 000 (1); and caregiver education level $>$ 12th grade (0) versus $\leq$ 12th grade (1). The cutoff for annual family income was chosen to best approximate those above versus below the poverty line for a family of four with two children (\$24,036 according to 2015 census data). The education level cutoff was chosen following guidelines set by previous CR research (e.g., Brody et al., 2013). The CR socioeconomic status (SES) variable is the sum of scores across these three dichotomized measures, with scores ranging from 0 to 3.

Parents gave consent for the research staff to access children's medical information, from which data were extracted about 1 year after the participant's study visit. Illness-related risk factors were aggregated into a single CR variable to demonstrate SCD disease

Table I. *Codes, Definitions, and Examples for Observed Macro and Micro Codes*

Code	Definition	Examples	*M* (*SD*)
Macro codes[a]			
Stimulates cognitive development	Parents' use of activities to enhance child's thinking, achievement, and learning in areas of perceptual, cognitive, and linguistic development.	Verbal: "See the rounded corners? Where is there a spot for that?"	4.68 (1.40)
Encourages Independence	Parental demonstrations of trust in and encouragement of the child's independence in thought and actions.	Verbal: "I know you can do it, just give it another try"	3.15 (1.72)
Listener responsiveness	The parent's nonverbal and verbal responsiveness as a listener to the verbalizations of the child through behaviors that validate and indicate attentiveness to the child.	Nonverbal: Eye contact, head-nods when child is speaking Verbal: "Yeah, mm-hmm."	5.60 (1.14)
Communication	The parents' ability to neutrally or positively express his/her own point of view, needs, wants, etc. in a clear, appropriate way, and reasonable manner; and to demonstrate consideration of the child's point of view. The good communication promotes rather than inhibits exchange of information.	Verbal: "That is an interesting idea."	5.87 (0.95)
Sensitive/child-centered	Parents' responses to child are appropriate and based on the child's behavior and speech; they offer support and independence so child can experience mastery, success, pride, and effective self-regulatory skills.	Nonverbal: Managing activity pace, handing child next piece. Verbal: "You seem frustrated, what's wrong?"	5.04 (1.50)
Micro codes			
Maintains	Caregiver continues the topic of the preceding utterance and/or develops the general topic of the conversation.	Child: "This one looks like a tree." Parent: "What else could it be?"	70.89 (27.14)
Expansions	Caregiver repeats some or all of the child's utterance but also adds additional content.	Child: "This way." Parent: "You want to turn it this way?"	1.57 (1.33)

[a]Macro codes were used to create responsive parenting composite.

severity: no history (0) versus prior overt or covert stroke (1); no history (0) versus prior acute chest event (1); hemoglobin level > 9.45 g/dL (0) versus < 9.45 g/dL (1) at most recent clinic appointment; and no visits (0) versus 1 or more visits to the emergency department (1) within the last year. These risk factors and cutoffs were chosen based on guidelines from previous research assessing pediatric SCD disease severity with multiple indicators (Logan et al., 2002; van den Tweel et al., 2010) and availability of data in participants' medical records. Disease CR ranged from 0 to 4.

Disease-Related Parenting Stress

Caregivers completed the Pediatric Inventory for Parents (Streisand, Braniecki, Tercyak, & Kazak, 2001), a 42-item parent-report questionnaire to assess both frequency and difficulty of disease-related parenting stress. For each item, caregivers were instructed to use a 5-point Likert scale to indicate how often the event has occurred (1 = *never* to 5 = *very often*) and how difficult the event has been (1 = *not at all* to 5 = *extremely*) during the past 7 days. For the purpose of the present study, the 8-item Medical Care Difficulty scale was used to assess stress related to parenting a child with SCD. Examples include, "Helping my child with medical procedures; Making decisions about medical care or medicines." Internal consistency in the present sample was $\alpha = 0.96$.

Data Analytic Strategy

The sample of 48 children and 48 caregivers resulted in .80 power to detect moderate effect sizes ($r \geq .39$) using uncorrected p values. All statistical analyses were conducted using IBM SPSS Statistics (Version 25). Cognitive outcomes were assessed for normality, and FSIQ and WMI were negatively skewed. Means and standard deviations of cognitive function scores and observed parenting codes were calculated. Pearson correlations were calculated to assess the three preliminary hypotheses, using two-tailed tests of significance and $\alpha = .05$. Bonferroni corrections for familywise error were performed for the tests of each of the hypotheses. Linear multiple regression analyses were used to assess the primary hypothesis.

Results

Descriptive Statistics

Scores for FSIQ ($M = 91.56$, $SD = 10.98$), WMI ($M = 93.65$, $SD = 14.58$), and Reading Composite ($M = 93.98$, $SD = 14.43$) were all significantly below the normative means on these measures ($p < .01$);

Table II. *Bivariate Correlations Among Measures of Child Cognitive Function, Parenting, and Parent Stress*

	1	2	3	4	5	6	7	8	9
1. Child age	–								
2. FSIQ	−.17	–							
3. WMI	−.29*	.73***	–						
4. Reading composite	−.16	.73***	.67***	–					
5. Responsive parenting[a]	−.11	.12	.10	.23	–				
6. Maintains[b]	−.02	.09	−.04	.20	.40**	–			
7. Expansions[b]	.05	.35*	.34*	.29*	.25^{+}	.44**	–		
8. Parental medical care stress	−.28^{+}	−.23	−.40**	−.43**	−.33*	−.23	−.23	–	
9. CR SES	−.05	−.11	−.16	−.15	−.00	−.06	−.06	.30*	–
10. CR disease	.05	−.05	−.22	−.16	.09	.28^{+}	.24^{+}	−.02	.09

Note. FSIQ = full scale IQ; WMI = Working Memory Index; CR = cumulative risk; SES = socioeconomic status.
[a]Composite of macro codes.
[b]Micro code.
$^{+}p < .10$; $^{*}p < .05$; $^{**}p < .01$; $^{***}p < .001$.

though they were each within the average range. With regard to observed parenting codes, the macrolevel responsive parenting composite ranged from 14 to 34 ($M = 24.34$, $SD = 5.52$), the microlevel maintains code ranged from 7 to 133 ($M = 70.89$, $SD = 27.84$), and the microlevel expansions code ranged from 0 to 4 ($M = 1.57$, $SD = 1.33$).

Bivariate Correlational Analyses

Bivariate analyses examining associations of exposure to socioeconomic and disease risk to parent stress are presented in Table II. The first hypothesis was partially supported, as higher levels of parenting stress were significantly associated with greater parental socioeconomic disadvantage ($r = .30$, $p = .043$), but parenting stress was not significantly associated with children's disease severity. Bonferroni correction for familywise error reduced $p < .025$, meaning the association between parent stress and socioeconomic disadvantage was no longer significant. The second hypothesis also received partial support, as parenting stress was significantly related to less responsive parenting as measured by the macrolevel coding system ($r = -.33$, $p = .026$), but it was not significantly related to either the maintains or expansions codes based on the microcoding of parents' behavior. After correcting for familywise error ($p < .017$), the association between stress and responsive parenting was no longer significant. The third hypothesis also received partial support in the bivariate analyses, as parenting stress was related to children's lower WMI ($r = -.40$, $p = .006$) and Reading Composite ($r = -.43$, $p = .003$), and parents' use of expansions (reflecting and building on what the child says) was significantly correlated with higher child FSIQ ($r = .35$, $p = .017$), WMI ($r = .34$, $p = .021$), and Reading Composite ($r = .29$, $p = .049$). Only the association between parent stress and child reading was significant after correcting for familywise error ($p < .005$). No significant associations were found for the responsive parenting composite or parents' use of maintains with the measures of cognitive function or achievement.

Multiple Regression Analyses

Regression analyses were used to test the fourth hypothesis (Table III); that is, the prediction of children's cognitive function and achievement from socioeconomic and disease risk, disease-related parent stress, and indicators of observed parenting behaviors at the macrolevels and microlevels. The model predicting children's FSIQ was significant, accounting for 19% of the variance, and parents' use of expansions was the only variable that accounted for unique variance in children's FSIQ ($\beta = .39$, $p = .018$). The model for children's WMI was significant, account for 35% of the variance, and cumulative disease risk factors ($\beta = -.30$, $p = .033$), parental stress ($\beta = -.43$, $p = .006$), and parental use of expansions ($\beta = .34$, $p = .022$) were unique predictors. Finally, the model predicting children's Reading Composite was significant, accounting for 31% of the variance, and cumulative disease risk factors ($\beta = -.29$, $p = .048$) and parental stress ($\beta = -.40$, $p = .016$) were unique predictors, and parental use of expansions approached significance ($\beta = .27$, $p = .075$).

Discussion

Children with SCD are at significant risk for impairments in cognitive function and a range of disease and social-environmental factors may contribute to these difficulties. We examined the role of caregivers as a salient proximal factor impacting the course of cognitive development in offspring with SCD. Caregivers of children with SCD may be unique in their exposure to two distinct areas of hardship: socioeconomic disadvantage and chronic health problems in children. Accordingly, these caregivers may experience significant stress that impairs the ability to provide

Table III. *Summary of Linear Regression Analyses for Variables Predicting Child Cognitive Function*

Predictor	Full scale IQ				Working memory				Reading comprehension			
	β	*SE*	*F*	R^2	β	*SE*	*F*	R^2	β	*SE*	*F*	R^2
			1.79	.19			4.06***	.35			3.26**	.31
CR SES	.02	1.56			.08	1.82			.06	1.89		
CR disease	−.21	1.52			−.30**	1.79			−.29**	1.89		
Parental medical stress	−.16	.24			−.43***	.28			−.40**	.29		
Responsive parenting	−.07	.33			−.09	.38			.01	.39		
Expansions	.39**	1.32			.34**	1.54			.27*	1.59		

Note. CR = cumulative risk.
*$p < .10$; **$p < .05$; ***$p < .01$.

responsive, cognitively stimulating parenting. Thus, the purpose of the present study was to extend research on stress, parenting, and cognitive function in the context of pediatric SCD (Brown et al., 1993). This study is the first to integrate research on the combined association of socioeconomic and parent stress and to explore how responsive parenting is related to cognitive performance in children with SCD.

Consistent with the first hypothesis, we found that family socioeconomic disadvantage was significantly related to greater medical care related parenting stress; however, child disease severity was not related to parent stress. This suggests that the stress of socioeconomic disadvantage may present more challenges for parents of children with SCD than the stress associated with the disease itself. Surprisingly, socioeconomic disadvantage was not directly associated with children's cognitive function or Reading Composite in this sample. Previously, King et al. (2014) found that annual household income and maternal education were significantly related to WASI FSIQ in the largest sample of children and adolescents with SCD to assess this relation. The current study may have been underpowered to find this direct association, yet findings suggest that there could be an indirect relation between SES and cognitive function through parent stress and behaviors.

In support of the second hypothesis higher levels of parents' medical care related stress was associated with lower levels of macrolevel responsive parenting. Further, parents' levels of medical care related stress were associated with lower scores on measures of children's working memory and reading, and parents' use of expansions was correlated with higher scores for children's FSIQ, WMI, and Reading Composite. Thus, the elevated levels of stress experienced by parents of children with SCD are related to lower levels of responsive parenting that in turn, is related to more deficits in cognitive function. These findings suggest that there is a pathway from parental stress through parenting to children's cognitive function that should be examined in future longitudinal research.

Finally, we found that parents' use of expansions in their interactions with their children during the puzzle-solving task was a significant predictor of FSIQ and working memory and approached significance for reading, even after controlling for disease and SES related risk factors. Disease related CR and medical care related parenting stress were also significant predictors of children's working memory and reading in the multivariate models. All three models were statistically significant, accounting for 19% of the variance in FSIQ, 35% of the variance in working memory, and 31% of the variance in reading. This pattern suggests that measures of complex aspects of cognitive function and school achievement are more strongly associated with the medical and social environmental factors included in these analyses. The findings are consistent with previous models that include social or environmental disadvantages as potential causes of cognitive problems in SCD (e.g., Brown et al., 2000).

The current study used multivariate models to include disease related risk factors, broad socioeconomic risk factors, and proximal measures of responsive parenting as predictors of cognitive function in children SCD. The findings are consistent with findings from other studies of parents and typically developing children (Landry et al., 2012) and children from families faced with economic disadvantage (Doan & Evans, 2011). Further, this study examined the role of responsive parenting with children with SCD and highlights the importance of parenting along with and parental stress as possible targets for interventions to enhance cognitive development in this at-risk group of children (Compas et al., 2017).

Previous research has provided evidence that children and adolescents with SCD are at risk for significant cognitive deficits (Prussien et al., 2019), and interventions to improve or slow the decline in cognitive function in these children are a high priority. Biological interventions, such as hydroxyurea and chronic blood transfusions have been shown to reduce the risk of silent cerebral infarcts (DeBaun & Kirkham, 2016), which are significantly related to increased deficits and the current findings provide

further support for these interventions. However, relatively few studies have addressed environmental sources of risk and have typically used global indicators of family SES. Schatz et al. (2004) found that both hematocrit and SES were significant unique predictors of cognitive function, as was the interaction of hematocrit level and SES. And Tarazi et al. (2007) found that SES was significantly correlated with the memory/attention. The current study suggests that SES may be related to cognitive function in children with SCD through more proximal processes of responsive parenting.

The current study provides evidence for a second important target for intervention—decreasing levels of stress and improving responsive parenting skills for parents of children with SCD. Only one study to date has tested the effect of a parenting intervention in pediatric SCD. In a sample of infants and preschool children with SCD, Fields et al. (2016) found significant improvement in cognitive and expressive language after completing a home visitation intervention to improve parenting skills. The current findings underscore the importance of the development and testing of interventions to support parents of children with SCD and to enhance their skills in providing a responsive environment to support cognitive growth and development.

The current study had several strengths and weaknesses. First, the cross-sectional design of the study limits conclusions that can be made from the significant associations among variables, as many of the associations may be bidirectional (see Burlew, Evans, & Oler, 1989). Future research using longitudinal designs is a priority. Second, the moderate sample size constrained statistical power to be able to detect only medium to large effects, and some effects were no longer significant after Bonferroni correction. Future multisite research with larger samples will provide greater power to detect small to medium effects that may be clinically meaningful. Third, it will be important to compare these processes in children with other chronic health conditions to determine if they are unique to SCD or are reflected in other pediatric conditions (Compas et al., 2017). These limitations notwithstanding, the present study utilized a multimethod design with validated measures to replicate and expand on previous research to provide evidence for the relation between parent and child functioning in SCD.

In summary, the findings from the present study highlight the potential impact of parent functioning on cognitive development in pediatric SCD. Further, the findings highlight the need to develop targeted interventions for parents of children with SCD to decrease levels of stress and enhance parenting skills, including attentive listening and responsive questions and expansions, with the aim improving cognitive functioning in youth.

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