Family Functioning, Neurocognitive Functioning, and Behavior Problems in Children With Sickle Cell Disease

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Objective: To investigate the independent and combined contributions of neurocognitive and family functioning to mother-reported behavior problems in children with sickle cell disease (SCD) and evaluate the factor structure of the Family Environment Scale (FES) with African American families.

Method: The study sample included 289 children enrolled in the multisite Cooperative Study of Sickle Cell Disease. The study protocol included neuropsychological evaluation and brain magnetic resonance imaging (MRI) of the children, and mothers completed the Child Behavior Checklist and Family Environment Scale.

Results: With child and maternal demographic parameters controlled, conflicted family functioning, but not neurocognitive functioning, accounted for a significant portion of the variance in mother-reported behavior problems. The factor structure of the FES for families of children with SCD was found to be similar to that for other families.

Conclusions: Family functioning may be a salient target for fostering adaptation to chronic childhood illness.

Key words: family functioning; neurocognitive functioning.

Sickle cell disease (SCD) is a major health problem that affects one in 400 African Americans (Consensus Conference, 1987). Although advances in health care have increased life expectancy, children with SCD are at increased risk for developmental and behavioral problems.

The evidence regarding the psychological adjustment of children and adolescents with SCD is similar to that for children with other chronic illnesses. There is an increased risk for adjustment problems, but good adjustment is also possible (Thompson et al., 1994) and internalizing problems are more frequent than externalizing problems (Thompson, Gil, Burbach, Keith, & Kinney, 1993).

Studies are now seeking to identify the factors and processes associated with the within-group variability in adjustment for children with chronic

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illness. Increasingly, these studies are guided by conceptual models that view chronic illness in system-theory terms as a potential stressor requiring adaptation (Thompson & Gustafson, 1996). Models differ in component processes but reflect three broad dimensions: illness/condition parameters, including type, duration, and severity; child parameters, including age, gender, and coping methods; and social-ecological parameters, including socio-economic status (SES), parental adjustment, and family functioning (Wallander & Thompson, 1995).

In 1989, Kazak advocated for the inclusion of family systems perspectives as part of the chronic illness model, and now family functioning is the most frequently investigated social-ecological correlate of adjustment of children with chronic illness (Wallander & Thompson, 1995). In particular, low levels of family conflict and high levels of family support or cohesiveness have been associated with favorable adjustment of children with chronic illness (Lavigne & Faiер-Routman, 1993). Now, models are addressing the interaction of illness parameters and family system variables in adaptation to chronic childhood illness. For example, Rolland (1993) hypothesized that the three-way goodness-of-fit among the psychosocial demands of the illness, the psychosocial demands of the family’s life cycle stage, and the style of family functioning are prime determinants of adaptation.

Parents of children with SCD reported more family conflict, less organization, and more emphasis on control than parents of healthy children (Burlow, Evans, & Olen, 1989), and coping competency was found to be positively associated with family cohesion and organization for adolescent boys and girls and negatively with family conflict for girls (Hurtig & Park, 1989). However, the interaction of family processes with illness/condition processes in adaptation of children with SCD has yet to be investigated.

In addition to being at high risk for adjustment problems, children with SCD also are at increased risk for neurocognitive deficits and cognitive and academic functional impairment. Cerebrovascular accidents (CVAs), primarily infarction, are estimated to occur in 5%–10% of children with sickle cell disease (Balkaran et al., 1992; Powars, Wilson, Imbus, Pegelow, & Allen, 1978). Furthermore, 11%–20% of children with sickle cell anemia with no neurologic deficit have been shown to have cerebral infarction detected by radiographic techniques such as magnetic resonance imaging (MRI) (Hindmarsh, Brozovic, Brook, & Davies, 1987; Pavlakis et al., 1988). Functional problems most often have been identified in visual motor integration, attention and concentration, arithmetic, memory, and reading (Armstrong et al., 1996).

The Cooperative Study of Sickle Cell Disease (CSSCD), a national natural history study with patients from 15 clinical sites (Farber, Koshy, & Kinney, 1985; Gaston & Rosse, 1982), includes an investigation of the association between neuropsychological deficits, as reflected in MRI findings, and cognitive and academic functioning in children 6–12 years of age (Armstrong et al., 1996). In this study (n = 194), 4.6% of the children had a clinical history of CVA and central nervous system (CNS) abnormalities were identified on MRI in 17.9% of the children. The children with a history of CVA performed significantly poorer than children with silent infarcts or no MRI abnormality on a number of measures of neurocognitive and academic performance. Furthermore, children with silent infarcts on MRI performed significantly poorer than children with no MRI abnormality on tests of arithmetic, vocabulary, and visual-motor speed and coordination. This study indicates that children with SCD, even without clinical evidence of stroke, are at risk for frontal lobe impairment, neurocognitive deficits, and poor academic performance (Armstrong et al., 1996). This finding of functional impairment in children without evidence of CVAs also was reported by Brown et al. (1993) in their study of children with SCD who performed significantly poorer than a nondiseased sibling comparison group on measures of attention and reading.

It is now clear that children with SCD are at increased risk for behavioral problems and neurocognitive and academic deficits. In general, with children with chronic illness, behavioral problems are associated with family functioning characterized by low support or high conflict and with impairments of the CNS (Thompson & Gustafson, 1996). However, it is not yet clear how neurocognitive functioning and family functioning act together in the adjustment of children with SCD.

The primary purpose of this study was to determine the independent and combined contribution of family functioning and neurocognitive functioning to psychological adjustment of children with SCD. More specifically, we hypothesized that behavior problems will be associated with (1) poor neurocognitive functioning, reflected in positive MRI findings, and low levels of intellectual func-
slightly higher verbal scale IQ scores (89.62 vs. 85.81), \(F(1, 382) = 4.75, p < .03\), performance scale IQ scores (91.46 vs. 87.43), \(F(1, 382) = 6.15, p < .02\), and full scale IQ scores (89.65 vs. 85.08), \(F(1, 382) = 7.79, p < .01\).

**Biomedical Parameters**

In terms of genotype, 196 children (68%) were SS and 93 children (32%) were SC. Hemoglobin diagnoses were confirmed by the Centers for Disease Control (CDC) for 286 of the 289 children. The hematocrit level, obtained within one year of and closest to the time of the neuropsychological evaluation, was used and ranged from 16% to 40% with a mean of 26% and a standard deviation of 5%. Although there is considerable variability, sickle cell anemia (SS) is usually more severe than the other types of SCD. High hematocrit levels have been associated with high pain rate (Platt et al., 1991).

**Method**

**Participants**

Four hundred thirty-two patients with either HbSS or HbSC disease from 15 clinical sites were enrolled in Phase 2 of the CSSCD multicenter natural history study between September 1989 and June 1991. Of these, 430 children, who were 6 years of age or older at the time of enrollment or who turned 6 while on study, were eligible for neuropsychological and MRI studies. Of these, 392 children completed the neuropsychological evaluation. The current study sample included 289 of the 392 children with a child behavior checklist completed by mothers who were living with their child (boys = 151 [52%]; girls = 138 [48%]). At the time the mothers were interviewed, the children ranged in age from 5.9 to 15.5 years (mean: 8.3 ± 2.1 years, median: 7.4 years).

Maternal age ranged from 20 years to 56 years with a mean of 33.08 (SD = 6.40). In terms of maternal education, 77 (27%) had less than a high school education, 75 (26%) were high school graduates or obtained the general equivalency degree (GED), 35 (12%) had some technical training after high school, 84 (29%) had some college education, 8 (3%) were college graduates, and 9 (3%) completed an advanced degree after college.

In terms of marital status, 98 (34%) of the mothers were married living with their spouse, 66 (23%) were divorced, separated, or widowed, and 122 (43%) were never married.

The study sample (n = 289) did not differ significantly (chi square) from the nonparticipants (n = 143) in the proportion of genotype (SS vs. SC), gender, or marital status (n = 286 vs. 29) or mean (ANOVA: n = 289 vs. 101) hematocrit and hemoglobin levels, child age, math achievement score, and reading achievement score. The study sample had

A secondary purpose of this study was to determine whether the factor structure of the Family Environment Scale characterizes African American families with children with SCD similarly to other nonillness and homogeneous ethnic samples.

**Procedure**

Informed consent to participate was obtained from parents. Demographic and medical history data were obtained through questionnaires and review of the CSSCD medical records for each child. In addition, each child had a brain MRI without contrast and a neuropsychological evaluation and his or her mother completed questionnaires regarding child behavior and family functioning.

**Behavioral Problems.** Mothers completed the Child Behavior Checklist (CBCL; Achenbach & Edelbrock, 1983), which assesses the frequency of 112 problem behaviors on a 3-point scale (not true, somewhat or sometimes true, very true or often true). Three global T-scores were used in this study: Total Problem Behaviors, Internalizing Behaviors, and Externalizing Behaviors. We adopted a cutoff score above the 90th percentile (T = 63) as indicative of a clinically significant behavior problem (because the study began in 1989, the 1983 and not the 1991 norms were used.)

**Neuropsychological Evaluation.** The standardized test battery, individually administered by trained examiners under the supervision of a licensed psychologist, was obtained on 283 children. The Wechsler Intelligence Scale for Children-Revised (WISC-R; Wechsler, 1974) was used to assess intellectual functioning and yielded full scale (FSIQ), verbal scale (VSIQ), and performance scale (PSIQ) intelligence quotients. The Woodcock-Johnson-Revised
Tests of Achievement (WJ-R; Woodcock & Johnson, 1989) was used to assess academic achievement and yielded broad reading and broad math cluster scores.

Brain Magnetic Resonance Imaging (MRI). MRIs of the brain without contrast were performed in accordance with the standard practices at each institution with or without sedation, depending upon the age and cooperation of the child. Each patient's MRI was reviewed independently by two neuroradiologists without knowledge of the patient’s diagnosis or clinical history and by a third neuroradiologist if there were disagreements. The MRIs were obtained on 244 children within one year of the neuropsychological evaluation and were classified into one of four groups: normal (n = 192), clinically apparent cerebral infarction (n = 12), silent cerebral infarction (n = 33), and atrophy (n = 7) (see Armstrong et al., 1996 for a more detailed description of MRI classification procedures and criteria).

Family Functioning. Family functioning was assessed with the Family Environment Scale (FES; Moos & Moos, 1981). The FES, completed by mothers within one year of the child's neuropsychological evaluation, was obtained on 270 families. The FES is a 90-item true-false scale designed to measure the social environments of families; it has good reliability and validity and is widely used as a measure of family environment (Moos & Moos, 1981). It is composed of 10 subscales that cluster into three empirically derived higher-order components (Kronenberger & Thompson, 1990). The Supportive component consists of the Cohesion, Expressiveness, Independence, Active-Recreational Orientation, and Intellectual-Cultural Orientation subscales; it measures family mutual interest, concern, support, and activities across a wide domain. The Conflicted component is comprised of the Conflict, Cohesion (negative direction), and Organization (negative direction) subscales; it reflects a dimension of conflict lacking in organization and mutual support. The Controlling component is a combination of the Control, Achievement Orientation, Moral-Religious Emphasis, and Independence (negative direction) subscales; it represents the use of expectations and rules to control the family environment. The FES component scores are calculated by adding (or subtracting, for subscales with negative loadings) the T-scores of constituent subscales (see Kronenberger & Thompson, 1990).

The component structure of the FES has been replicated with two samples, adolescent and adult, drawn from the normative FES data (Kronenberger & Thompson, 1990). In terms of construct validity, children (n = 109) with chronic illness, including diabetes, cancer, and myelodysplasia, whose mothers reported behavior problems, had family functioning characterized by higher levels of conflict and lower levels of supportiveness than children without behavior problems (Kronenberger & Thompson, 1990).

Results

FES Component Structure

To determine whether the subscales of the FES cluster together into similar components for African American families with children with SCD as for other families, a principal components analysis of the T-scores for the ten FES subscales was performed (n = 270). Three components with eigenvalues > 1 were retained and subjected to promax rotation.¹

The component structure of the FES of children with SCD closely replicated previous findings (Kronenberger & Thompson, 1990). The coefficients of congruence (Gorsuch, 1973) with the chronic childhood illness, adult normative, and adolescent normative samples respectively were Supportive component (.92, .92, .86); Conflicted component (−.90, −.87, −.84); and Controlling component (.85, .86, .90). Because of this high degree of congruence, established scoring rules for FES components (Kronenberger & Thompson, 1990) were applied to the current sample.

Behavior Problems

With the criterion of T > 63, the percentage of children with behavior problems was 25% in terms of Total Behavior Problems, 22% for Internalizing Behavior Problems, and 18.0% for Externalizing Behavior Problems. Overall, 30% (86/289) of the children had a mother-reported behavior problem (Total or Internalizing or Externalizing Behavior Problem). There were no significant differences (chi-square) in the rate of behavior problems as a function of gender (male, 30%; female, 29%) or MRI status (normal, 28%; clinical, 25%; silent, 27%). There was no significant difference in the rate of

¹A more extensive report of the finding of the FES factor analysis, including a table presenting factor loading, is available upon request.
behavior problems of children with SS (22%) and children with other types of SCD (30%). Children whose mothers were never married had significantly higher \( \chi^2(2, n = 258) = 6.667, p = .04 \), rates of externalizing behavior problems (25%) than children whose mother was divorced, separated, or widowed (12%) and children whose parents were married and living together (13%).

The relationship between behavior problems and child and maternal parameters and family processes was examined in several ways. First, Pearson zero-order correlations were obtained and are presented in Table I. There were significant, but weak, negative correlations of total and externalizing behavior problem scores with maternal education and the neurocognitive parameters of intellectual and academic functioning. In contrast, there were moderately strong and significant correlations of behavior problem scores with dimensions of family functioning. The Supportive factor was negatively correlated and the Conflicted factor was positively correlated with externalizing, internalizing, and total behavior problem scores. The Controlling factor was significantly positively correlated with only the externalizing behavior problem score.

Second, behavior problem subgroups were formed on the basis of having a total, internalizing, or externalizing behavior problem score in the clinical range (T \( > 63 \)) and subgroup differences in variables were examined by ANOVA. The findings presented in Table II indicate that the subgroup with, compared to the subgroup without, behavior problems (based on total behavior problem score) had significantly lower verbal scale IQ scores, lower reading and math scores, and family functioning characterized by lower levels of support and higher levels of conflict. These analyses were also conducted with behavior problem subgroups formed on the basis of externalizing and internalizing scores. The same general pattern of findings was demonstrated with the exception that the subgroup with, compared to the subgroup without, externalizing behavior problems did not differ significantly on the math cluster and were significantly higher in FES controlling in addition to other FES factors. The subgroup with, compared to the subgroup without, internalizing behavior problems did not differ significantly in IQ scores.

Hierarchical multiple regression analysis was used to assess the unique and combined contribution of variables to total, internalizing, and externalizing behavior problem scores, respectively. The order of entry among sets of variables was determined to reflect the increment in variance in behavior problems accounted for by the incorporation of neurocognitive and family functioning variables over and above that accounted for by the demographic and biomedical parameters. Thus, child age and gender and maternal age, education, and marital status (married vs. other) and the biomedical parameters of genotype and hematocrit were controlled by being forced to enter first. Because there was no a priori basis for setting the order of entry for the neuropsychological and family

### Table I. Pearson Correlations between CBCL Behavior Problem Scale Scores and Child and Mother Variables

<table>
<thead>
<tr>
<th>Variables</th>
<th>N</th>
<th>Total</th>
<th>Externalizing</th>
<th>Internalizing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child age</td>
<td>289</td>
<td>-0.01</td>
<td>0.00</td>
<td>-0.01</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>283</td>
<td>0.08</td>
<td>0.12*</td>
<td>0.02</td>
</tr>
<tr>
<td>VSIQ</td>
<td>283</td>
<td>-0.15**</td>
<td>-0.17**</td>
<td>-0.12</td>
</tr>
<tr>
<td>PSIQ</td>
<td>283</td>
<td>-0.12*</td>
<td>-0.13*</td>
<td>-0.09</td>
</tr>
<tr>
<td>FSIQ</td>
<td>283</td>
<td>-0.16**</td>
<td>-0.17**</td>
<td>-0.11</td>
</tr>
<tr>
<td>Reading cluster</td>
<td>276</td>
<td>-0.15*</td>
<td>-0.14*</td>
<td>-0.12*</td>
</tr>
<tr>
<td>Math cluster</td>
<td>272</td>
<td>-0.15*</td>
<td>-0.13*</td>
<td>-0.15*</td>
</tr>
<tr>
<td>Mother age</td>
<td>289</td>
<td>-0.00</td>
<td>-0.00</td>
<td>-0.00</td>
</tr>
<tr>
<td>Mother education</td>
<td>288</td>
<td>-0.12*</td>
<td>-0.14*</td>
<td>-0.08</td>
</tr>
<tr>
<td>Supportive</td>
<td>270</td>
<td>-0.27***</td>
<td>-0.32***</td>
<td>-0.17***</td>
</tr>
<tr>
<td>Conflicted</td>
<td>270</td>
<td>0.42***</td>
<td>0.44***</td>
<td>0.29***</td>
</tr>
<tr>
<td>Controlling</td>
<td>270</td>
<td>0.11</td>
<td>0.16**</td>
<td>0.03</td>
</tr>
</tbody>
</table>

* \( p < .05 \), ** \( p < .01 \), *** \( p < .001 \).

### Table II. Comparison of Subgroups with and without Behavior Problems on Child and Mother Variables

<table>
<thead>
<tr>
<th>Variables</th>
<th>Behavior problems ( n = 86 )</th>
<th>No behavior problems ( n = 203 )</th>
<th>ANOVA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Child age</td>
<td>7.81 2.00</td>
<td>7.94 2.08</td>
<td>1,287 0.23</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>25.97 5.56</td>
<td>26.18 5.26</td>
<td>1,281 0.09</td>
</tr>
<tr>
<td>VSIQ</td>
<td>86.39 14.59</td>
<td>90.97 14.51</td>
<td>1,281 5.82*</td>
</tr>
<tr>
<td>PSIQ</td>
<td>90.19 13.74</td>
<td>91.99 14.23</td>
<td>1,281 0.95</td>
</tr>
<tr>
<td>FSIQ</td>
<td>87.18 13.45</td>
<td>90.68 13.88</td>
<td>1,281 3.80</td>
</tr>
<tr>
<td>Reading cluster</td>
<td>88.21 17.71</td>
<td>94.65 18.34</td>
<td>1,274 7.35**</td>
</tr>
<tr>
<td>Math cluster</td>
<td>88.11 17.13</td>
<td>95.67 18.12</td>
<td>1,270 10.37**</td>
</tr>
<tr>
<td>Mother age (yrs)</td>
<td>32.72 6.58</td>
<td>33.23 6.33</td>
<td>1,287 0.38</td>
</tr>
<tr>
<td>Supportive</td>
<td>240.75 39.97</td>
<td>257.37 32.89</td>
<td>1,268 12.61**</td>
</tr>
<tr>
<td>Conflicted</td>
<td>-51.96 28.86</td>
<td>-69.74 19.49</td>
<td>1,268 34.65***</td>
</tr>
<tr>
<td>Controlling</td>
<td>127.79 19.07</td>
<td>125.40 18.38</td>
<td>1,268 0.93</td>
</tr>
</tbody>
</table>

* \( p < .05 \), ** \( p < .01 \), *** \( p < .001 \).
functioning parameters, a forward stepwise procedure was used. Thus, the neuropsychological parameters of verbal, performance, and full scale IQ; reading and math cluster scores; and MRI status (normal vs. other, i.e., clinical infarction, silent infarction, and atrophy); and the three family functioning factors were allowed to enter if they accounted for a significant ($p < .05$) increment in behavior problem variance.

The demographic and biomedical parameters together accounted for 2%–6% in total, internalizing, and externalizing behavior problem scores. The MRI status and neuropsychological parameters did not account for a significant portion of variance in any of the behavior problem scores. However, conflicted family functioning level accounted for a significant 19% increment in variance in total behavior problem scores, 11% in internalizing behavior problem scores, and 20% in externalizing behavior problem scores. Table III presents the summary of the multiple regression analysis for the Total Behavior Problem score. The pattern was similar for internalizing and externalizing behavior problems.

### Discussion

The primary hypotheses of this study, that behavior problems would be associated with neurocognitive and family functioning, received mixed support. In terms of the association of neurocognitive parameters and behavior problems, the zero-order correlations were significant but not very strong. Those with a behavior problem, compared to those without, had significant and moderately lower verbal IQ scores and reading and math scores. Although the association of positive MRI findings and lower cognitive and academic functioning has been demonstrated previously (Armstrong et al., 1996), there was no significant relationship between MRI findings (normal, clinical, silent) and frequency of behavior problems. One possible explanation is that there were too few children with clinical evidence of a cerebrovascular accident ($n = 12$) to adequately assess the relationship of MRI status to behavior problems.

In terms of family functioning, both the zero-order correlations and the behavior problems subgroup contrasts indicated a significant and moderately strong relationship of behavior problems to lower levels of the Supportive component and higher levels of the Conflicted component. This pattern of association of maternal perceptions of family functioning and mother-reported behavior problems is consistent with previous findings with children with other chronic illnesses (Kronenberger & Thompson, 1990, see Lavigne & Fraier-Routman, 1993; Thompson & Gustafson, 1996; Wallander & Thompson, 1995).

The hierarchical multiple regression analysis did not provide support for the hypothesis of independent and combined contributions of neurocognitive and family functioning to behavior problems. With child and maternal demographic parameters controlled, only Conflicted family functioning accounted for a significant increment in the amount of variance accounted for in total, internalizing, and externalizing behavior problems.

Taken as a whole, the findings of this study indicate that family functioning, more specifically family functioning characterized as conflicted, and not neurocognitive functioning, is related to mother-reported behavior problems in children with SCD. The Conflicted component reflects high levels of conflict and a lack of both organization and mutual support. These findings are consistent with those showing that family factors, in addition to other social-ecological parameters such as maternal anxiety and depression (Thompson et al., 1993), influence mothers’ perceptions and report of behavior problems in their children with SCD.

These findings also have potential clinical implications in regard to fostering adaptation to chronic childhood illness. That is, family functioning characterized as conflicted could be evaluated as a

<p>| Table III. Hierarchical Multiple Regression Analysis of Total Behavior Problem Score |</p>
<table>
<thead>
<tr>
<th>Variables</th>
<th>$\beta$</th>
<th>$R^2$</th>
<th>Cumulative $R^2$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Demographics</td>
<td>0.02</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Child age</td>
<td></td>
<td>-0.04</td>
<td></td>
</tr>
<tr>
<td>Child gender</td>
<td></td>
<td>-0.03</td>
<td></td>
</tr>
<tr>
<td>Mother age</td>
<td></td>
<td>-0.01</td>
<td></td>
</tr>
<tr>
<td>Mother education</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mother’s marital status</td>
<td></td>
<td>-0.08</td>
<td></td>
</tr>
<tr>
<td>Biomedical parameters</td>
<td>0.02</td>
<td>0.04</td>
<td></td>
</tr>
<tr>
<td>Genotype</td>
<td></td>
<td>-0.11</td>
<td></td>
</tr>
<tr>
<td>Hematocrit</td>
<td></td>
<td>-0.04</td>
<td></td>
</tr>
<tr>
<td>Family functioning</td>
<td>0.19</td>
<td>0.23</td>
<td></td>
</tr>
<tr>
<td>Conflicted factor</td>
<td>0.45</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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marker of families requiring more extensive psychosocial assessment and support. Furthermore, family functioning could be a salient intervention target. Now we need studies at the experimental level of research (Thompson & Gustafson, 1996), that is, intervention studies that endeavor to improve adaptation of children with chronic illness by increasing family supportiveness and organization and reducing family conflict. Experimental level research is needed to confirm theories and correlational evidence of the role of family functioning in adaptation to chronic illness.

An important consideration pertains to the appropriateness of various constructs and measures with African American families. With regard to the measure of children's behavior problems, the normative group for the CBCL was chosen to be representative of the population of the United States on SES, ethnicity, region, and urban-suburban-rural residence, and there were minimal differences in ratings by parents of different ethnic groups matched for SES (Achenbach, 1991). Although the SCD sample was not matched to the CBCL norms, it is not inappropriate to use the CBCL with this sample. Furthermore, the resulting frequency of behavior problems was approximately 2.5 times the rate for the CBCL normative group, which is consistent with findings across a number of illness subgroups (see Thompson & Gustafson, 1996).

Similarly, an important component of this study was the replication of the factor structure of the FES with African American families of children with SCD. Furthermore, the family functioning factors related to adjustment in the same patterns with African American families as with other families. That is, mother-reported behavior problems were associated with lower levels of family supportiveness and higher levels of family conflict.

There are several limitations of this study. First, the exclusive reliance on maternal report for assessment of behavior problems means that a differentiation cannot be made between maternal perceptions of child behavior and actual child behavior. Moreover, it is also well established that mother's own psychological adjustment affects her perceptions of her children's behavior (see Thompson & Gustafson, 1996). Furthermore, relying upon the report of a single observer, the mother, in a study of family systems is not ideal, particularly when the single observer is assessing both family functioning and behavior problems. These, of course, are well-recognized problems (see Thompson & Gustafson, 1996) that persist because of the cost and complexity of obtaining reports from other informants. Again, the best method of disentangling these effects is to move to the experimental level of research after there is consistent evidence across studies of the association of family functioning with mother-reported children's behavior problems.

Acknowledgments

This work was supported by the Division of Blood Diseases and Resources of the National Heart, Lung, and Blood Institute of the National Institutes of Health.


Received February 5, 1998; accepted September 28, 1998
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