Validity of the Pediatric Quality of Life Inventory for Youth with Sickle Cell Disease

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Objective  Evaluate the validity of the Pediatric Quality of Life Inventory (PedsQL) for sickle cell disease (SCD).

Methods  Sixty-eight parent–child dyads (children 5–18 years) completed the PedsQL. Medical record review assessed history of specific morbidities.

Results  Internal consistency of the scales varied. The strongest reliability was for parent proxy-report for specific domains or for global functioning scores with either informant. Modest internal consistency was found for specific domains with child informants, particularly for younger children. Moderate convergent validity was found between informants. History of neurologic problems or major pain episodes indicated criterion validity for specific scales.

Conclusions  The PedsQL appears to validly assess quality of life in youth with SCD. Domain-specific measurement of quality of life was limited by (a) low reliability for youth-report and (b) lack of discriminant validity. Choice of informant may be important when evaluating quality of life effects from pain or neurologic problems in SCD.

Key words  disease complications; pediatric; quality of life; sickle cell disease.

Sickle cell disease (SCD) is a set of inherited blood disorders that can result in a host of physiological, cognitive, and psychosocial comorbidities, including chronic and acute anemia, infection, stroke, severe pain episodes, delayed puberty, and academic underachievement (Midance & Shand, 1992; Schatz & Puffer, 2006; Shapiro, 1993). These complications are frequently unpredictable and can lead to disruptions in the daily lives of children with SCD as well as their ability to fulfill major life roles. For example, pain in youth with SCD results in elevated rates of school absenteeism, which in turn can lead to poor school performance and interruptions in peer relations (Fuggle, Shand, Gill, & Davies, 1996; Shapiro et al., 1995). In other illness groups, such as spinal bifida, pain has been linked to lower quality of life and higher rates of depression (Oddson, Clancy, & McGrath, 2006). Current research indicates inconsistent findings regarding the rates of clinically significant anxiety and depression in youth with SCD; however, feelings of hopelessness and low self-esteem following frequent hospitalizations, pain, and school absenteeism are commonly reported (Anie, 2005). Neurologic complications, including silent and overt stroke can result in deficits in attention and concentration skills as well as academic and occupational concerns for the majority of persons with these complications (Armstrong et al., 1996; Hariman, Griffith, Hurttig, & Keehn, 1991; Schatz, Brown, Pascual, Hsu, & DeBaun, 2001; Wang et al., 2001).

Life expectancy of individuals with SCD in the USA has improved dramatically from the early 1970s when the median life span was ~14 years to the early 1990s when the median life span ranged between the mid-40s and late 60s depending on subtype and gender (Platt et al., 1994). With improvements in life expectancy for individuals with SCD, the importance of quality of life has increased. Practitioners and researchers seek additional ways to understand disease impact and to evaluate treatment outcomes beyond traditional morbidity and mortality outcome measures, such as stroke incidence and frequency of pain episodes. The impact of a health condition on a persons’ physical, psychosocial, and role functioning has been referred to as Health-related Quality of Life (HRQOL; Pal, 1996). Self-reports of HRQOL are of particular importance in evaluating disease and treatment effects, as they allow
for a subjective perspective of HRQOL that goes beyond objective measures of mortality and morbidity.

Previous research has found links between parent proxy-report and child self-report of HRQOL and that these relationships are moderated by aspects of SCD, such as disease complications such as history of acute chest syndrome, frequency of pain episodes, and neurologic comorbidities (Panepinto, O’Mahar, DeBaun, Loberiza, & Scott, 2005). This and other research has demonstrated that factors unique to SCD, including greater number of disease complications, frequency of pain episodes, and neurologic comorbidities are associated with decrements in HRQOL as assessed by the Child Health Questionnaire (CHQ; Palermo, Schwartz, Drotar, & McGowan, 2002; Panepinto et al., 2005). However, attempts to examine such relationships with other HRQOL tools have not found such links (Barakat, Lutz, Smith-Whitley, & Ohene-Frempong, 2005), suggesting the possible important of one’s choice in measurement methods for accurately assessing HRQOL.

Evaluating the reliability of parent-reported HRQOL is of particular importance in pediatric SCD, as children with SCD experience complications, such as pain episodes and strokes that can reduce their ability to communicate effectively with others on a short- and long-term basis. Parents routinely serve as proxy-reporters on formal and informal assessments of their child’s HRQOL, yet few researchers have compared parent and child ratings of HRQOL in pediatric SCD (Barakat et al., 2005; Panepinto et al., 2005). Of this work, only Panepinto and colleagues conducted a statistical comparison of parent and child ratings across the 10 subscales of the CHQ. Their investigation included children with SCD ages 5–18 years. Results indicated moderate and statistically significant correlations (.33–.58) on observable domains, such as physical limitations resulting from pain experience and lower correlations (.10–.25) on domains that are not readily observable by parents, such as self-esteem and social well-being (Panepinto et al., 2005). This work, as well as research with other illness groups, such as cancer, has shown that parents often rate their children as more limited than does the child themselves (Levi & Drotar, 1999; Panepinto et al., 2005).

The current investigation seeks to assess the validity of the PedsQL, a generic HRQOL tool previously found to be reliable and valid for use with other pediatric conditions including pediatric cancer, diabetes, and obesity (Schwimmer, Burwinkle, & Varni, 2003; Varni et al., 2003; Varni, Burwinkle, Katz, Meeske, & Dickinson, 2002). The PedsQL has several potential advantages over previously employed measures of HRQOL in youth with SCD, such as the CHQ. Specifically, the PedsQL has parallel forms for parent proxy-report and youth self-report that allows for direct comparisons between parent and child report by domain and individual items for children ranging between 5 and 18 years of age (Varni, Seid, Knight, Uzark, & Szer, 2002). This measure is both brief and comprehensive, allowing for parent and youth ratings on school functioning in addition to other key domains. Given the link between SCD and academic difficulties, assessing school functioning is essential to the evaluation of disease-related impacts on HRQOL (Schatz, 2004; Schatz et al., 2001). Although the CHQ includes an assessment of school functioning on the 87-item child-report and 50-item parent proxy-report versions, the short-form versions do not include this domain. With only 23-items, the PedsQL may be more feasible to use as part of routine clinical care. As a generic HRQOL instrument, the PedsQL also allows for potential comparisons between different illness groups.

The goal of the present report was to assess the validity of the PedsQL in youth with SCD by evaluating the reliability of its scales, construct validity, and criterion validity. Reliability was assessed by evaluating the internal consistency of parent proxy-report and youth self-report for discrete domains as well as the instrument’s summary scores. There is considerable evidence that quality of life can be conceptualized as a domain-specific construct (Power, Harper, & Bullinger, 1999); however, summary scores are a more concise way to quantify HRQOL in situations in which there are not meaningful differences across specific domains. Given the broad age range of youths included, we examined internal consistency and construct validity in younger and older age grouping to assess potential age-related impact on reliability and validity. Construct validity was evaluated by investigating convergent and discriminant validity between and within raters and scales using a Multi-Trait Multi-Method (MTMM) Matrix. The validity of using parents as proxy reporters for children’s HRQOL was assessed by evaluating cross-informant correspondence for the same scale. Finally, the criterion validity of the PedsQL in assessing HRQOL was investigated by evaluating its relationship to two predominant sources of morbidity in individuals with SCD: neurologic complications and episodes of moderate-to-severe pain. It was hypothesized that the PedsQL in the SCD population would provide internally consistent scores, show moderate correspondence across informants, and yield lower scores for HRQOL in children with either neurobehavioral morbidity or a history of major pain episodes.
Methods
Participants
Medical chart reviews were conducted for 84 children with SCD between the ages of 5 and 18 years ($M = 12$ years, $SD = 3$ years) who participated in a psychosocial screening as part of their routine care between September 2004 and September 2006 at a Pediatric Hematology/Oncology outpatient clinic in the southeastern USA. Participation in annual psychosocial screening at the clinic was strongly encouraged by the treating hematologist and considered a part of routine care. Of the patients who did not participate in the screening due to time constraints, the large majority were screened as part of their next appointment. No patients refused to participate in the screening; however, data was not kept on the rate of participants who were unable to participate on multiple occasions or who did not receive the screening because they did not return to the clinic. These screenings include a semi-structured interview, completion of the PedsQL to assess the child’s well-being, and a measure of caregiver stress to screen for parent risk. Among the 84 potential cases, 68 had completed both parent and youth self-reports. Twelve cases were missing youth self-report and four cases were missing parent proxy-report.

All of the participants were identified in the medical records as African-American. Fifty percent were females. Most were insured through the state Medicaid program (54%) with a minority of patients having private insurance (28%) or both Medicaid and private insurance (18%). Patient’s were most frequently accompanied by their mothers (84%) followed by fathers (10%) and other guardians (6%). The SCD subtypes were 68% HbSS, 15% HbSC, 10% HbSβ+, and 7% HbSβ0. The mean hematocrit value for this sample for the routine blood draw closest to completion of the study measures was 28.6% ($SD = 5.3$). These values indicate relatively lower hematocrit when compared to the normative values, which range between 33% and 49% depending on child age and gender. Nine of the children had a history of overt stroke and six of these children were receiving chronic blood transfusion therapy at the time the study measures were collected.

Measures
PedsQL
Parent proxy-report and youth self-report of HRQOL was obtained using the fourth edition of the PedsQL (Varni, Seid et al., 2002). The PedsQL parent proxy-report and youth self-report form includes 23 items assessing a child’s functioning in Physical (eight items), Emotional (five items), Social (five items) and School (five items) domains. Respondents indicated how much of a problem the child has had over the previous 1 month with specific aspects of functioning on a 0–4 scale, with 0 indicating “never a problem” and 4 indicating “almost always a problem”. The responses for each item are reverse scored and linearly transformed to a 0–100 scale with higher scores reflecting better HRQOL. Domain scores are obtained by summing the items and dividing by the number of items answered per domain to account for missing data. If >50% of the items for any domain are missing a scale score was not calculated. The Psychosocial Health score was derived by computing the mean of the sum of the item scores of the Emotional, Social, and School domains. A Total Score was also computed by obtaining the average of the domain scores.

Medical Chart Review
Details of patient’s disease complications, comorbidities, and demographics were obtained through medical chart review using a medical chart checklist to provide a structured form for the review of paper and electronic medical charts. The first author completed the chart review for all patients and the second author independently reviewed 44% of the same charts. Kappa coefficients for the two key variables for the study, history of neurobehavioral morbidity, or a history of major pain episodes was .64 and .85, respectively, indicating “good” to “very good” inter-rater reliability (Fleiss, 1981). For data analyses, any disagreements on coding were resolved through discussion of the criteria by the two coders. If a disagreement on definitions was resolved in favor of the secondary coder, then all the charts of patients were reviewed for that variable to maintain consistency in definitions. Demographic information included patient age, gender, ethnicity, and insurance funding source. Illness-specific information included disease subtype, transfusion status, neurobehavioral concerns (history of stroke, silent cerebral infarcts, moyamoya disease, meningitis, seizure disorder, or formally diagnosed disorders in learning, language, or developmental disorder), and history of major pain episodes over the previous 2 years. A 2-year period of time was selected instead of a 1-year period because it fully captures the pain history for patients with a significant but unpredictable pain. Disorders in learning, language, or development were considered present if patient had supporting documentation, such as neuropsychological assessment confirming the diagnosis, reports of outside evaluators, or a parent had reported to the hematologist that their child had an individualized education plan to address learning concerns. Other neurobehavioral concern were considered to be present if found in documentation.
by the hematologist, other medical specialists, or by MRI/MRA results. Major pain episodes included SCD-related pain episode that required hospitalization or Emergency Department care as documented in the hematologist notes, discharge documents, or documentation from treating facilities.

**Procedure**

Appropriate institutional board approval was obtained prior to medical chart review. The first author reviewed the electronic and paper charts of patients who had participated in a screening. Both types of charts were reviewed to ensure the inclusion of documents from other treating facilities that may have not been included in electronic charts. Screenings were conducted on an annual basis as part of the patient’s routine Hematological health maintenance appointments. Following their physical examination, patients were accompanied by the hematologist to the psychologist’s office located within the clinic.

The psychologist (C.B.M.) explained to all new patients that the goal of the screening was to check on all aspects of a patient’s well being and to introduce the psychosocial services available through the clinic. Parents and patients were initially queried as to any current concerns in areas of school and home behavior and general family life. Next, the PedsQL was introduced as a tool to provide a better understanding of the patient’s functioning in their daily life. The psychologist reviewed the PedsQL instructions and demonstrated how to complete a few sample items to ensure parent comprehension. Parents completed the measure in the nearby waiting room, while the psychologist assisted the patient in completing the measure in the psychologist’s office. This method was employed both to ensure comprehension of all items and as a clinical tool to facilitate inquiry into areas in which the patient indicated difficulties. Researchers comparing the self versus interviewer-administrated versions of the PedsQL reported no significant differences in scores in youth with brain tumors or cerebral palsy (Palmer, Meeske, Katz, Burwinkle, & Varni, 2007; Varni et al., 2005).

**Data Analysis**

Less than 1% of the parent proxy-report and youth self-report data contained missing data. Per the test developer’s instructions for the instrument, subscale mean imputation was employed to obtain domain scores for participants with missing data.

Age groups were formed by splitting the groups into a child group (<13 years) and an adolescent group (>13 years), allowing for the examination of possible reporting differences between children and adolescents. To assess reliability Cronbach’s $\alpha$ was computed for each informant and PedsQL scale as an evaluation of the internal consistency for all scales. To assess convergent validity, Pearson correlations were computed between parent proxy-report and youth self-report. Intraclass correlations (ICC) were also computed across informants, in order to obtain an index of absolute agreement in light of between-subject and total variability. Correlations for the Physical Health and Psychosocial Health summary scores were also examined in the correlation matrix for different informant–different scale correlations and same informant–different scale correlations to describe discriminant validity. The summary scores were used for the full MTMM matrix due to questionable reliability of specific psychosocial domains for child-report.

For descriptive purposes, the level of agreement across informants was compared to other pediatric populations. The Pearson cross-informant correlations were compared to those of a recent evaluation of parent proxy-report in children with cancer using the PedsQL (Varni, Burwinkle et al., 2002) to determine whether parent proxy-report for SCD was similar to what is found in the other illness groups. Scores across parent proxy-report and youth self-report were also compared with paired samples $t$-tests to assess whether parents were systematically providing ratings higher or lower than youths for each domain and for global scores. A set of within-subjects $t$-tests were conducted to describe the profile of PedsQL scores for Physical Health, Emotional Functioning, Social Functioning, and School Functioning by determining scales with relatively higher versus lower ratings. Finally, between-group $t$-tests were calculated comparing the self and proxy-report total scores from the current sample to a population-based estimate of African-American youths (Varni, Burwinkle, Seid, & Skarr, 2003).

To assess criterion validity children with neurobehavioral morbidity or history of major pain were identified as criterion groups. The $t$-tests were used to compare ratings for each criterion group to the children without each of the morbidities. The $\alpha$-level for all tests was set at .05.

**Results**

**Internal Consistency**

The Cronbach’s $\alpha$ data are presented in Table I. Parent proxy-report yielded values of .8 or higher for all scales except for School Functioning. Youth self-report tended to yield somewhat lower values with the exception of the internal consistency of the Total Score and Psychosocial Health summary score. These lower values appeared to be
due predominantly to the younger children providing lower internal consistency scores for their self-ratings than older children. The School Functioning scale showed the lowest internal consistency across both informants. Examination of the item inter-correlations indicated two sets of items for School Functioning. There was moderate inter-correlation among items 1, 2, and 3 (items related to paying attention, forgetfulness, and keeping up with schoolwork) with a Cronbach α coefficients of .68 for parent proxy-report and .51 for self-report. Items 4 and 5 (items related to missing school due to illness or medical appointments) demonstrated stronger correlations with a Cronbach α of .71 for parent proxy-report and .79 for self-report. Thus, the school functioning scale appeared to have at least two dimensions within this sample.

**Construct Validity**

Due to relatively low internal consistency of the specific psychosocial domain scores for youth self-report, cross-informant correspondence for the Physical and Psychosocial Health summary scores was the primary focus for assessing construct validity (Table II). These Pearson correlation values were statistically significant (p < .05) for all comparisons except for reports of Physical Health by the younger children in comparison to their parents, supporting the idea of convergent validity through cross-informant correspondence in most comparisons. Varni, Burwinkle et al. (2002) reported cross-informant correlations for a group of 190 pediatric oncology patients with values of .57 for Physical Health and .49 for Psychosocial Health. The cross-informant correlation for our SCD sample is lower for Physical Health (p < .01) than that reported by Varni, but not statistically different for Psychosocial Health (p = .56). The absolute agreement between parent proxy-report and youth self-report based on ICC demonstrated a similar pattern of statistically significant convergence across informants with a correlation of .22 (p < .05) for Physical Health and .29 (p < .01) for Psychosocial Health for the entire sample.

Inspection of the MTMM matrix across constructs (“traits”) and informants (“methods”) suggested a probable methods factor. The absolute values of the same informant–different construct correlations tended to be higher than the same construct–different informant values (Tables II–IV), which is contrary to the data pattern indicating discriminant validity for the constructs. Different informant–different construct correlations are also supposed to be the lowest values in the matrix, but our data consistently did not yield lower correlation values than that for different informant–same construct comparisons.

Although we did not have sufficient statistical power to provide a strong direct statistical test of differences between these correlations, overall, the data did not suggest discriminant validity for Physical versus
Psychosocial Health. Highly similar patterns of correlations were also found in MTMM matrices for specific psychosocial domains, indicating more evidence for a “methods” factor than for a “trait” factor in the pattern of correlations (data not shown). The Pearson correlation across parent proxy-report and child self-report for Total Score was \( r(68) = .41, p < .001, \) for the total sample, \( r(36) = .32, p < .05, \) for children less than 13 years, and \( r(32) = .52, p < .01, \) for children 13 years and older.

**Criterion Validity**

Thirty-five children had some history of neurologic or neurobehavioral morbidity and 45 of the children had a history of a major pain episode over the past 24 months. A Fischer’s exact test comparing group membership across neurologic and pain history suggested these two types of morbidity were largely independent of each other in this sample \( (p > .7). \)

PedsQL ratings for neurologic history and major pain history subgroups are shown in Table III. The most notable pattern to these findings was related to informant-based differences. There were predominant decrements in HRQOL for the children with neurologic morbidity according to parent proxy-report including lower Total Score, Psychosocial Health, Emotional Functioning, and School Functioning. In the group with a history of major pain, parents reported lower School Functioning when compared to youth without a history of major pain.

### Table III. Mean (SD) Pediatric Quality of Life Inventory Scores

<table>
<thead>
<tr>
<th>Scale</th>
<th>Positive history (n = 35)</th>
<th>No history (n = 33)</th>
<th>(95% CI)</th>
<th>d</th>
<th>Positive history (n = 45)</th>
<th>No history (n = 23)</th>
<th>(95% CI)</th>
<th>d</th>
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<tbody>
<tr>
<td><strong>Neurologic morbidity subgroups</strong></td>
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<tr>
<td>Total score</td>
<td>64.3 (18.1)</td>
<td>67.4 (16.4)</td>
<td>(−5.35 to 11.43)</td>
<td>−0.18</td>
<td>62.7* (17.5)</td>
<td>71.9* (15.3)</td>
<td>(0.51 to 17.73)</td>
<td>−0.56</td>
</tr>
<tr>
<td>Physical health</td>
<td>67.8 (21.5)</td>
<td>71.0 (18.1)</td>
<td>(−6.39 to 12.87)</td>
<td>−0.16</td>
<td>65.9* (21.0)</td>
<td>76.1* (15.5)</td>
<td>(0.29 to 20.09)</td>
<td>−0.55</td>
</tr>
<tr>
<td>Psychosocial health</td>
<td>62.8 (18.4)</td>
<td>66.2 (17.9)</td>
<td>(−5.55 to 12.19)</td>
<td>−0.19</td>
<td>61.4* (18.3)</td>
<td>70.4* (16.7)</td>
<td>(−0.04 to 18.18)</td>
<td>−0.51</td>
</tr>
<tr>
<td>Emotional functioning</td>
<td>66.6 (24.2)</td>
<td>64.4 (20.0)</td>
<td>(−12.95 to 8.59)</td>
<td>0.09</td>
<td>63.9 (23.5)</td>
<td>68.7 (19.1)</td>
<td>(−6.52 to 16.14)</td>
<td>−0.22</td>
</tr>
<tr>
<td>Social functioning</td>
<td>69.4 (21.2)</td>
<td>72.6 (26.3)</td>
<td>(−8.38 to 14.69)</td>
<td>−0.13</td>
<td>66.8* (24.7)</td>
<td>79.1* (19.5)</td>
<td>(0.53 to 24.18)</td>
<td>−0.55</td>
</tr>
<tr>
<td>School functioning</td>
<td>52.9* (19.5)</td>
<td>61.5* (15.7)</td>
<td>(−0.09 to 17.24)</td>
<td>−0.49</td>
<td>53.9* (16.6)</td>
<td>63.5* (19.6)</td>
<td>(0.53 to 18.70)</td>
<td>−0.53</td>
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<tr>
<td><strong>Major pain subgroups</strong></td>
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<tr>
<td>Total score</td>
<td>60.5* (16.0)</td>
<td>67.0* (14.6)</td>
<td>(−0.90 to 13.81)</td>
<td>−0.42</td>
<td>61.6 (15.0)</td>
<td>67.7 (15.8)</td>
<td>(−1.77 to 13.86)</td>
<td>−0.39</td>
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<tr>
<td>Physical health</td>
<td>60.5 (20.1)</td>
<td>62.1 (19.5)</td>
<td>(−7.93 to 11.28)</td>
<td>−0.08</td>
<td>59.1 (20.4)</td>
<td>65.5 (17.9)</td>
<td>(−3.55 to 16.51)</td>
<td>−0.33</td>
</tr>
<tr>
<td>Psychosocial health</td>
<td>60.6* (16.6)</td>
<td>68.7* (15.7)</td>
<td>(0.34 to 15.99)</td>
<td>−0.50</td>
<td>62.5 (15.9)</td>
<td>68.6 (17.4)</td>
<td>(−2.34 to 14.48)</td>
<td>−0.37</td>
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<tr>
<td>Emotional functioning</td>
<td>64.6* (23.5)</td>
<td>73.6* (14.8)</td>
<td>(−0.50 to 18.63)</td>
<td>−0.46</td>
<td>66.8 (20.4)</td>
<td>73.3 (19.3)</td>
<td>(−3.77 to 16.74)</td>
<td>−0.33</td>
</tr>
<tr>
<td>Social functioning</td>
<td>66.3 (22.3)</td>
<td>71.4 (21.9)</td>
<td>(−5.64 to 15.79)</td>
<td>−0.23</td>
<td>68.0 (21.9)</td>
<td>70.2 (23.0)</td>
<td>(−9.16 to 13.59)</td>
<td>−0.10</td>
</tr>
<tr>
<td>School functioning</td>
<td>50.9** (15.4)</td>
<td>61.2** (18.3)</td>
<td>(2.18 to 18.53)</td>
<td>−0.61</td>
<td>52.7* (16.0)</td>
<td>62.2* (19.0)</td>
<td>(0.77 to 18.24)</td>
<td>−0.54</td>
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</tbody>
</table>

Major pain groups were based on history of a major pain episode over the past 2 years.

*Scores in the row differ between groups according to t-tests \( p < .05. \)

**Scores in the row differ between groups according to t-tests \( p < .01. \)

**Comparisons of Inter-rater and Inter-domain Mean Scores**

The t-tests comparing scores for the Physical Health scale showed lower parent proxy-report than youth self-report scores, \( t(67) = 2.87, p < .01, \) (Table IV). The remaining scales showed no statistical differences between youth and proxy report.

Pairwise comparisons of scales to assess profiles of HRQOL ratings are shown in Table IV. School Functioning was rated significantly lower than Physical Health, Emotional Functioning, and Social Functioning across both informants. The profile comparison for youth self-report showed children rated Physical Health, \( t(67) = 6.08, \) \( p < .01, \) Social Functioning, \( t(66) = 5.61, \) \( p < .01, \) and Emotional Functioning \( t(66) = 3.40, \) \( p < .01, \) significantly higher than the School Functioning. Parents rated Physical Health \( t(67) = 2.11, \) \( p < .05, \) Social Functioning \( t(67) = 5.54, \) \( p < .01, \) and Emotional Functioning \( t(67) = 5.20, \) \( p < .01, \) significantly higher than School Functioning. Youths also rated their Emotional Functioning significantly lower than their Social Functioning, \( t(67) = 5.61, \) \( p < .01. \) In contrast, parents rated Emotional Functioning, \( t(67) = 2.88, \) \( p < .01, \) and Social Functioning, \( t(67) = 2.84, \) \( p < .01, \) significantly higher than Physical Health.
convergent validity was supported, discriminant validity across domains appeared to be poor, suggesting limited evidence for domain-specific measurement in our study. Both the internal consistency data and the lack of evidence supporting discriminant validity suggest the PedsQL may be best suited for measuring overall HRQOL in SCD rather than assessing discrete domains. This measurement pattern for the PedsQL may reflect either limitations of the instrument or the underlying nature of HRQOL decrements in SCD.

Our assessment of criterion groups supported informant-specific criterion validity for the PedsQL. Parents of youth with neurobehavioral complications reported lower HRQOL relative to youth without these complications for Total Score and several specific domains. Interestingly, comparing the youth self-report of these two groups revealed differences in school functioning only. In contrast, youth with a history of pain episodes reported generally lower HRQOL than those without a history of major pain episodes, whereas parents reported decrements for school functioning only. These data pattern is consistent with concerns raised by other researchers about the validity of youth self-report for HRQOL in children with neurologic disease (Panepinto et al., 2005). The data also suggest parent proxy-report may not be as sensitive as youth self-report to pain-related effects on functioning in school-age children.

These criterion validity results add to the literature by demonstrating the importance of considering both informant and SCD complications on ratings of HRQOL. The impact of neurological complications, such as poor attention span or emotional distress, on the daily functioning of youth with SCD may be best reported by caregivers who can compare their child’s functioning to that of other youth. Due to its subjective and personal nature, the impact of pain on functioning may be less accessible to parents and thus lend itself better to self-reported assessments of functioning. Researchers have reported on similar discrepancies in other illness groups and concluded that parent and child report of HRQOL may be based on different data, and thus should be viewed as complementary to each other but not equivalent (Jokovic, Locke, & Guyatt, 2004).

Comparisons of the current parent proxy and youth self-report PedsQL scores to those found in a population-estimate sample of African-American youths indicated statistically lower HRQOL in patients with SCD. Although this comparison group is matched on race/ethnicity alone and does not take into account the impact of other important variables, such as socioeconomic status, the data

### Table IV. Mean (SD) PedsQL Scores for Entire Sample

<table>
<thead>
<tr>
<th>Scale</th>
<th>Entire sample (n = 68)</th>
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<tbody>
<tr>
<td><strong>Self-report</strong></td>
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<tr>
<td>Physical health</td>
<td>69.3, (19.8)</td>
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<tr>
<td>Emotional functioning</td>
<td>65.5, (22.1)</td>
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<tr>
<td>Social functioning</td>
<td>71.2, (23.8)</td>
</tr>
<tr>
<td>School functioning</td>
<td>57.2, (18.1)</td>
</tr>
<tr>
<td>Psychosocial health</td>
<td>64.5, (18.1)</td>
</tr>
<tr>
<td>Total score</td>
<td>65.8 (17.3)</td>
</tr>
<tr>
<td><strong>Proxy-report</strong></td>
<td></td>
</tr>
<tr>
<td>Physical health</td>
<td>61.3a, (19.7)</td>
</tr>
<tr>
<td>Emotional functioning</td>
<td>69.0a, (20.1)</td>
</tr>
<tr>
<td>Social functioning</td>
<td>68.8a, (22.1)</td>
</tr>
<tr>
<td>School functioning</td>
<td>55.9a, (17.3)</td>
</tr>
<tr>
<td>Psychosocial health</td>
<td>64.5a, (16.6)</td>
</tr>
<tr>
<td>Total score</td>
<td>63.7 (13.4)</td>
</tr>
</tbody>
</table>

*Within informant comparison across specific scales are reported to provide a profile analysis. Scores that do not share a subscript letter differ significantly by paired sample’s t-test (p < .05). Total score and psychosocial summary scores were not included in paired samples t-tests.*

The t-tests comparing Total Scores of the current sample to those found in a population-based African-American sample indicated significantly lower scores for both child self-report \( t(209) = 7.49, p < .01 \), and parent proxy-report \( t(305) = 9.74, p < .01 \) in the SCD sample. These values represent effect sizes of Cohen’s \( d = 1.04 \) for child self-report and \( d = 1.12 \) for parent proxy-report.

### Discussion

Parent proxy-report and youth self-report on the PedsQL were compared to evaluate the validity of this tool to assess HRQOL in the pediatric SCD population. Convergent validity was demonstrated in that parent proxy-report and youth self-report on the PedsQL were significantly correlated, indicating that parents can serve as proxy reporters for youth HRQOL. These correlations appeared to be larger than those reported for other tools of HRQOL in pediatric SCD (Panepinto et al., 2005). Although there are no widely agreed upon standards for internal consistency, in clinical psychology a minimum Cronbach’s \( \alpha \) of near .9 is often used as a standard for instruments used in making decisions about an individual with values of \( \geq .7 \) recommended for research purposes (Clarke & Watson, 1995; Groth-Marnat, 2003). The PedsQL demonstrated adequate internal consistency for clinical purposes, as well as good reliability at the level of Total Score. For research purposes, parent proxy-report of specific domains and summary scores typically reached adequate internal consistency. While
suggest a relatively large impact of SCD on HRQOL in this population with effect sizes of $d > 1.0$.

A number of the current findings are consistent with those found in research investigating HRQOL in other pediatric illness groups. The cross-information correlations were similar to those found in research investigating the PedsQL in a pediatric oncology sample for the Psychosocial Health Summary score (Varni, Burwinkle et al., 2002). Domain-specific internal consistency was similar to that reported by other researchers (Varni, Sied, & Kurtin, 2001). The breakdown of the school functioning domain into two dimensions was consistent with the pattern found in pediatric populations with acute and chronic health conditions (Varni et al., 2001). Parents rated their child’s physical functioning as more limited than did youth themselves (Levi & Drotar, 1999; Panepinto et al., 2005). Finally, as was found in youth with spinal bifida, recent pain was associated with lower HRQOL Total Score on the PedsQL (Oddson et al., 2006).

The intra-item correlation breakdown for school functioning suggests that this domain may be comprised of two dimensions, one being school difficulties related to paying attention and completing schoolwork and the other being difficulties stemming from frequent school absences. Research has shown a pattern, whereby school absences can predict academic attainment but not necessarily achievement, whereas cognitive ability can predict both achievement and attainment (Schatz, 2004). Neurocognitive deficits and school absenteeism may represent two distinct sources of school problems for youth with SCD and other pediatric health conditions. Different types of school difficulties may require different types of interventions. Distinguishing between these sources of school problems may help researchers and clinicians to better refine the assessment of HRQOL and consider specific types of intervention needed to address school concerns in youth with SCD.

The results of this research must be interpreted within the limitations of the study. Compared to those treated at the local health department, youth treated at the hospital-based clinic where participants were recruited typically experience higher rates of neurological complications and pain morbidity, and are more likely to be above income guidelines for public insurance. The rates of disease-related complications in our sample were generally comparable to those reported by other researchers investigating HRQOL in pediatric SCD in children’s hospitals and academic medical centers (Palermo et al., 2002; Panepinto et al., 2005), suggesting that our findings will generalize to patients treated in these settings. For this preliminary work, we grouped participants by history of neurological complications and a significant pain history; however, there exists great variability within these groupings and our approach may have limited our statistical power to detect a relationship between disease morbidity and HRQOL. We did not believe the retrospective medical chart review would provide reliable estimates of small individual differences in the severity of neurologic or pain morbidity to measure these complications on a meaningful continuum of severity. There are inherent limitations with the use of retrospective data from patient’s medical records. While every attempt was made to obtain all documents from other healthcare providers and to thoroughly review the available records, some disease complications relevant to this study may not have been documented. Further, the medical chart review did not include a measure of SCD pain that was treated outside of the healthcare settings, limiting our ability to fully evaluate the role of pain in HRQOL. Finally, the sample size used in the current work was relatively small. It is noteworthy that most of the null effects for our criterion group comparisons (Table III) were small effect sizes using conventions described by Cohen (1988), whereas the statistically significant findings were approximately of medium size. Thus, future research is needed to cross validate the current findings with other larger samples predominantly to replicate patterns and increase the generalizability of the findings.

The use of a brief tool to routinely assess HRQOL is important to evaluate clinical and research outcomes. The PedsQL is a brief generic measure of HRQOL that is widely used in a range of illness groups. Additional work is needed to establish how the PedsQL compares to other measures of HRQOL. Our research generally supports the use of summary scores from the PedsQL parent proxy and youth self-report for evaluating HRQOL in the pediatric SCD population; however, the overall Total Score probably has the strongest validity evidence. In clinical practice, we have also found that routine clinical screenings that include a brief semi-structured interview and the PedsQL to obtain parent proxy-report and youth self-report has significant clinical utility. The goal of the screenings is to monitor the child’s functioning in home and school settings and to make appropriate referrals when specific areas of concern are identified, for example, referrals for school advocacy or psychoeducational assessment when school functioning concerns are identified. The PedsQL often reveals parent concerns that are not initially raised during unstructured queries about their child’s functioning. Additionally, using the self-report PedsQL facilitates discussion between the clinician and patient about the
patient’s functioning and highlights the patient’s greatest areas of concern through numerical ratings. Thus, we have found the specific items and domains to have clinical utility in the process of identifying concerns at an individual patient level. The present study further demonstrates the importance of obtaining both parent and child report when assessing HRQOL, especially in patients with a history of neurological and pain histories. Other researchers have found that parent characteristics, such as education level and marital status, moderate parent-reported HRQOL for youth with SCD or cancer (Panepinto et al., 2005; Sawyer, Antioniou, Toogood, & Rice, 1999). Additional work is needed to further evaluate family and patient characteristics, including a prospective accounting of disease severity, degree of caregiver stress, and available family resources that may moderate HRQOL ratings. Longitudinal work is needed to identify factors that may protect or preserve HRQOL in light of significant health concerns. This work will allow us to determine the best targets for interventions to improve HRQOL.

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Conflicts of interest: None declared.

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