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PedsQL™ Sickle Cell Disease Module: Feasibility, Reliability and Validity

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Abstract

Background—Sickle cell disease (SCD) is an inherited chronic disease that is characterized by complications such as recurrent painful vaso-occlusive events that require frequent hospitalizations and contribute to early mortality. The objective of the study was to report on the initial measurement properties of the new PedsQL™ SCD Module for pediatric patient self-report ages 5-18 years and parent proxy-report for ages 2-18 years.

Procedure—The 43-item PedsQL™ SCD Module was completed in a multisite study by 243 pediatric patients with SCD and 313 parents. Participants also completed the PedsQL™ 4.0 Generic Core Scales and PedsQL™ Multidimensional Fatigue Scale.

Results—The PedsQL™ SCD Module Scales evidenced excellent feasibility, excellent reliability for the Total Scale Scores (patient self-report $\alpha = 0.95$; parent proxy-report $\alpha = 0.97$),

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Conflict of Interest Statement

Dr. Varni holds the copyright and the trademark for the PedsQL™ and receives financial compensation from the Mapi Research Trust, which is a nonprofit research institute that charges distribution fees to for-profit companies that use the Pediatric Quality of Life Inventory™.

and good reliability for the nine individual scales (patient self-report α = 0.69-0.90; parent proxy-report α = 0.83-0.97). Intercorrelations with the PedsQL™ Generic Core Scales and PedsQL™ Multidimensional Fatigue Scales were medium (0.30) to large (0.50) range, supporting construct validity. PedsQL™ SCD Module Scale Scores were generally worse for patients with severe versus mild disease. Confirmatory factor analysis demonstrated an acceptable to excellent model fit.

Conclusions—The PedsQL™ SCD Module demonstrated acceptable measurement properties. The PedsQL™ SCD Module may be utilized in the evaluation of SCD-specific health-related quality of life in clinical research and practice. In conjunction with the PedsQL™ Generic Core Scales and the PedsQL™ Multidimensional Fatigue Scale, the PedsQL™ SCD Module will facilitate the understanding of the health and well-being of children with SCD.

Keywords

Sickle Cell Disease; PedsQL; pediatrics; children; health-related quality of life; patient-reported outcomes

Introduction

Sickle cell disease (SCD) is an inherited chronic disease characterized by complications such as recurrent painful vaso-occlusive events that require frequent hospitalizations. Prior work using generic health-related quality of life (HRQOL) instruments have demonstrated that patients with SCD experience significantly impaired HRQOL in their baseline health that worsens during acute complications. [1-4]. Generic HRQOLinstruments allow comparison of populations with different diseases or to healthy patients, but are limited when evaluating disease-specific functioning. Disease-specific HRQOL instruments are designed to evaluate functioning specific to a particular disease and are better able to detect differences within a population of patients. To the best of our knowledge, a validated pediatric SCD-specific HRQOL instrument does not exist in the empirical literature. In order to better understand differences in health status within the population of children with SCD and to enhance the ability to measure the impact of disease modifying therapies from the patient's and parent's perspectives, we developed the PedsQL™ SCD Module to address this significant gap in the literature [5].

Understanding the basic measurement properties of this disease-specific HRQOL instrument is critical prior to utilizing it in clinical trials and clinical practice. The objective of this study was to determine the initial measurement properties for the child self-report and parent proxy-report versions of the new PedsQL™ SCD Module, including feasibility, reliability and validity. We hypothesized that children with more severe SCD would have worse HRQOL than those with mild disease as measured by the PedsQL™ SCD Module, and that the SCD-specific scales would be significantly associated with generic HRQOL and fatigue.

Methods

Study Population and Procedures

Data collection place between June, 2010 and August, 2012. Participants were children ages 5-18 years and parents of children ages 2-18 years with a physician confirmed diagnosis of SCD (any genotype) at five clinical centers across the United States (Medical College of Wisconsin/Children's Hospital; University of Texas Southwestern/Children's Medical Center, Dallas; Baylor College of Medicine/Texas Children's Hospital, Houston; Jonathan Jaques Children's Cancer Center/Miller Children's Hospital, Long Beach, CA; University of Alabama at Birmingham/Children's of Alabama). The study population includes a convenience sample of eligible patients and/or parents who presented for a clinic visit. Children known to the clinical team to have cognitive impairment that would prevent them from understanding questions on the instrument were excluded. The human subjects institutional review boards at each center approved the study.

Measures

The PedsQL™ Sickle Cell Disease Module—The PedsQL™ SCD Module was developed through a literature review of relevant research, consultation with SCD experts, focus interviews, cognitive interviews, and pre-testing protocols [5]. Development of the items for the PedsQL™ SCD Module began in May, 2008 [5]. The child self-report items are listed in Supplemental Appendix I.

The 43-item PedsQL™ SCD Module > encompasses nine scales: 1) Pain and Hurt (9 items), 2) Pain Impact (10 items), 3) Pain Management and Control (2 items), 4) Worry I (5 items), 5) Worry II (2 items), 6) Emotions (2 items), 7) Treatment (7 items), 8) Communication I (3 items), 9) Communication II (3 items). The format, instructions, Likert response scale, and scoring method for the PedsQL™ SCD Module are identical to the PedsQL™ 4.0 Generic Core Scales, with higher scores indicating better HRQOL and lower SCD symptoms/problems [6].

The Module Scales are comprised of parallel child self-report and parent proxy-report formats for children ages 5-18 years, and a parent proxy-report format for children ages 2-4 years. Child self-report forms are specific for ages 5-7, 8-12, and 13-18 years. Parent proxy-report forms are specific for children ages 2-4 (toddler), 5-7 (young child), 8-12 (child), and 13-18 (adolescent), and assess parents' perceptions of their child's HRQOL. The instructions ask how much of a problem each item has been during the past month. The grammar and syntax of the new items were structurally equivalent to those in the existing PedsQL™ item bank. Instructions and response scales for the PedsQL™ SCD Module were created to be consistent with that of the PedsQL™ 4.0 Generic Core Scales for ages 2-18 years and PedsQL™ Disease-Specific Modules [6-11]. The PedsQL™ 5-point Likert-type response scale has been widely utilized in studies (0 = never a problem; 1 = almost never a problem; 2 = sometimes a problem; 3 = often a problem; 4 = almost always a problem), and has undergone extensive cognitive interviewing for many pediatric Patient Reported Outcomes Measurement Information System (PROMIS) scales and was found acceptable and understood by patients and parents [12-14]. To increase ease of use for the young child

self-report (ages 5-7), the response scale is simplified to a 3-point scale (0 = not at all a problem; 2 = sometimes a problem; 4 = a lot of a problem). This is consistent with the PedsQL™ 4.0 Generic Core Scales and the PedsQL disease-specific modules [6-11]. Forms are self-administered by the parent or child ages 8-18 years. For children 5-7 years of age, forms are interviewer-administered [6].

Items are reverse-scored and linearly transformed to a 0-100 scale (0=100, 1=75, 2=50, 3=25, 4=0), so that higher scores indicate better HRQOL. Scale Scores are computed as the sum of the items divided by the number of items answered (this accounts for missing data). If more than 50% of the items in the scale are missing, the scale score is not computed. This accounts for the differences in sample sizes for scales reported in the Tables. Although there are other strategies for imputing missing values, this is consistent with previous PedsQL™ publications and other well-established HRQOL measures [6,15,16]. To create the PedsQL™ SCD Module Total Scale Score (43 items), the mean is computed as the sum of the items divided by the number of items answered.

The PedsQL™ 4.0 Generic Core Scales—The 23-item PedsQL™ Generic Core Scales encompass: 1) Physical Functioning (8 items), 2) Emotional Functioning (5 items), 3) Social Functioning (5 items), and 4) School Functioning (5 items) [6]. The Physical Health Summary Score is the same as the Physical Functioning Scale. To create the Psychosocial Health Summary Score, the mean is computed as the sum of the items divided by the number of items answered in the Emotional, Social, and School Functioning Scales. The Generic Core Scales are scored similarly to the SCD Module scales. The PedsQL™ Generic Core Scales have demonstrated reliability, validity, and responsiveness in SCD [1,17-21].

The PedsQL™ Multidimensional Fatigue Scale—The 18-item PedsQL™ Multidimensional Fatigue Scale encompasses three domains including General Fatigue (6 items), Sleep/Rest Fatigue (6 items) and Cognitive Fatigue (6 items)[7,22]. The PedsQL™ Multidimensional Fatigue Scale has been validated among samples of children with numerous chronic health conditions [7,8,22-26], including SCD [18]. The scales are scored similarly to the SCD Module scales, with higher scores indicating better HRQOL (lower fatigue).

The PedsQL™ Family Information Form—Parents completed a modified PedsQL™ Family Information Form which contains demographic information including the child's date of birth, gender, race/ethnicity, and parental education [6].

Statistical Analysis

Feasibility was determined from the percentage of missing values [16]. Cronbach's coefficient alpha was utilized to determine scale internal consistency reliability [27]. Scales with internal consistency reliabilities of 0.70 or greater are recommended for comparing patient groups, while an internal consistency reliability criterion of 0.90 is recommended for analyzing individual patient scores [28]. Range of measurement was based on the percentage of scores at the extremes of the scaling range, that is, the maximum possible score (ceiling effect) and the minimum possible score (floor effect). Surveys with small floor or ceiling

effects (1% to 15%) are considered to meet acceptable measurement standards, while surveys with moderate floor or ceiling effects (> 15%) are considered less precise in measuring latent constructs at the extremes of the scale [29].

Factor analysis was conducted on the a priori hypothesized factor structure of the SCD Module. Since the purpose was to test an a priori hypothesized factor structure, confirmatory factor analysis (CFA) was utilized [30]. Given the commonly cited limitations of the chi-square statistic [31-33], and consistent with recommendations set forth in the literature [32-35], we examined different model indices of practical fit including the Root Mean Squared Error of Approximation (RMSEA) [34], the Comparative Fit Index (CFI) [32], the Tucker-Lewis Index (also known as the Non-Normed Fit Index; NNFI) [35], and the Normed Fit Index (NFI) [36]. Excellent model fit is suggested by RMSEA values ≤ 0.06, while acceptable model fit is suggested by RMSEA values between 0.06 and 0.08 [37,38]. For the NFI, NNFI and CFI indices, excellent model fit is suggested by values greater than or equal to 0.95, while acceptable model fit is suggested by values between 0.90 and 0.95 [32,36,39]. LISREL for Windows program was utilized for these analyses [40].

The sensitivity of a measurement instrument may be demonstrated through a cross-sectional analysis of differences between groups of patients with varying degrees of disease severity[41]. Sensitivity of the PedsQL™ SCD Module was determined by comparing patients with severe and mild SCD using independent samples *t*-tests. Disease status was classified a priori as mild or severe disease regardless of the child's SCD type which is consistent with prior work [17,21,42]. Patients were classified as having severe disease if they experienced one or more of the following complications of SCD: 1) overt stroke, 2) acute chest syndrome, 3) 3 or more hospitalizations for painful events in the prior 3 years. This classification was based on the criteria used for intervention with hydroxyurea or bone marrow transplantation [43-45]. All others were classified as having mild disease. We hypothesized that the SCD Module Scales would distinguish between mild and severe disease. Effect sizes were calculated to determine the magnitude of the differences [46]. Effect size as utilized in these analyses was calculated by taking the differences between the severe and mild SCD sample means, divided by the pooled standard deviation. Effect sizes for differences in means are designated as small (0.20), medium (0.50), and large (0.80) in magnitude [46].

An analysis of the intercorrelations among the PedsQL™ Generic Core Scales and the PedsQL™ Multidimensional Fatigue Scales with the SCD Module Scales was used to further examine construct validity of the PedsQL™ SCD Module. Computing the intercorrelations among scales provides initial information on the construct validity of an instrument [47]. We hypothesized that greater disease-specific symptoms/problems would correlate with lower overall generic HRQOL as measured by the PedsQL™ Generic Core Scales and the PedsQL™ Multidimensional Fatigue Scale based on the conceptualization of disease-specific symptoms as causal indicators of generic HRQOL [48]. Pearson Product Moment Correlation coefficients effect sizes are designated as small (0.10), medium (0.30), and large (0.50) [46].

Intraclass Correlation Coefficients (ICCs) were used to determine agreement between patient self-report and parent proxy-report [49]. The ICC provides an index of absolute agreement as it takes into account the ratio between subject variability and total variability [49,50]. ICCs are designated as ≤0.40 poor to fair agreement, 0.41-0.60 moderate agreement, 0.61-0.80 good agreement, and 0.81-1.00 excellent agreement [51,52]. Statistical analyses were conducted using SPSS Version 19.0 for Windows [53].

Results

Study Population

A total of 321 families participated (243 children ages 5-18 years and 313 parents/caregivers of children ages 2-18 years; 8 children completed the Module without parent/caregiver completion). The average age of the 153 males (47.7%) and 168 females (52.3%) was 9.66 years (SD = 4.85). With respect to race/ethnicity, the sample contained 315 (98.1%) self-reported Black non-Hispanic, 2 (0.6%) White non-Hispanic, 2 (0.6%) Hispanic, and 2 (0.6%) Other. With respect to parent education (n=321), 19.0% of mothers and 14.6% of fathers did not complete high school; 18.4% of mothers and 18.4% of fathers had a high school degree; 35.2% of mothers and 20.2% of fathers completed some college; 14.3% of mothers and 6.9% of fathers had a college degree; and 3.7% of mothers and 5.0% of fathers had a graduate degree (missing: 9.3% mothers and 34.8% fathers). The distribution of participants by site is as follows: Medical College of Wisconsin/Children's Hospital n=150; University of Texas Southwestern/Children's Medical Center, n=100; Baylor College of Medicine/Texas Children's Hospital, n=23; Jonathan Jaques Children's Cancer Center/ Miller Children's Hospital, Long Beach, n=26; University of Alabama at Birmingham/ Children's of Alabama n=22.

Feasibility: Missing Item Responses

The percentage of missing item responses was 3.0% and 2.6% for the SCD Module self-reports and proxy-reports, respectively and were distributed relatively equally across scales. For child self-report and parent proxy-report on the PedsQL™ Generic Core Scales, missing item responses were 2.7% and 3.8%, respectively, for all scales except the parent proxy-report School Functioning Scale. Missing items for the proxy-report School Functioning Scale were 5.3% (ages 5-18) and 13.4% (ages 2-4). This large percentage for toddlers may exist since instructions on the PedsQL™ toddler form ask parents to complete the School Functioning Scale if their child attends school or daycare and many toddlers do not attend school or daycare. Missing item responses on the PedsQL™ Multidimensional Fatigue Scale were 2.0% and 3.7% for child self-reports and parent proxy-reports, respectively.

Range of Measurement

Table I contains the percentage of scores at the extremes of the scaling range (floor and ceiling effects) for the PedsQL™ SCD Module Scales. For child self-report, there were no significant floor effects for any of the scales, and a ceiling effect for the Worry II, Emotions, and Communication I Scales. For parent proxy-report, there were no significant floor effects for any of the scales, and ceiling effects for the Pain Management and Control, Worry I, Worry II, Emotions, Treatment, Communication I, and Communication II Scales.

Internal Consistency Reliability

Internal consistency reliability coefficients for the PedsQL™ SCD Module are shown in Table I. All child self-report and parent proxy-report scales on the SCD Module exceed the minimum reliability standard of 0.70 required for group comparisons, except for the 2-item Emotions Scale on child self-report (0.69). The Total Scale Scores for both child self-report and parent proxy-report exceed the reliability criterion of 0.90 recommended for analyzing individual patient scores.

Confirmatory Factor Analysis

The results demonstrated that the 43 items of both the self-report and proxy-report versions loaded on nine latent variables consistent with the a priori hypothesized factor structure. For self-report, the goodness of fit statistics were RMSEA=0.068, NFI=0.90, CFI=0.96 and NNFI=0.95. For proxy-report, the fit statistics were RMSEA=0.079, NFI=0.94, CFI=0.97 and NNFI=0.96. These results demonstrated an acceptable to excellent model fit.

Sensitivity

Table II presents the differences between the pediatric patients with severe and mild SCD for the PedsQL™ SCD Module. For the Total Score, Pain and Hurt, Pain Impact and Emotions Scales, pediatric patients with severe SCD reported statistically significant lower disease-HRQOL than those with mild disease. For all parent proxy-report scales, except for the Communication I Scale, parents of pediatric patients with severe SCD reported statistically significant lower disease-HRQOL than those with mild disease.

Table III presents the intercorrelations between the PedsQL™ SCD Scales and Total Scale Score with the Generic Core Scales and summary scores. The majority of the intercorrelations are in the medium to large effect size range, supporting construct validity of the SCD Scales and Total Scale Score for child self-report and parent proxy-report.

Table IV presents the intercorrelations between the PedsQL™ SCD Scales and Total Scale Score with the Multidimensional Fatigue Scale and Total Scale Score. The majority of the intercorrelations are in the medium to large range effect size range, supporting construct validity of the SCD Scales and Total Scale Score for child self-report and parent proxy-report.

Parent/Child Agreement

ICCs between child and parent report for the PedsQL™ SCD Module Scales are shown in Table V. The majority of the ICCs are in the poor to fair and moderate agreement range.

Discussion

These findings support the initial feasibility, reliability and validity of the PedsQL™ SCD Module Scales and Total Scale Score in pediatric patients with SCD. The PedsQL™ SCD Module scales should be a useful multidimensional SCD-specific instrument that can be integrated with the PedsQL™ Generic Core Scales and the PedsQL™ Multidimensional

Fatigue Scale to provide a comprehensive assessment of patient generic and disease-specific HRQOL in pediatric patients with SCD.

The Module Scales were feasible with minimal missing values, supporting the notion that patients ages 8-18 and parents were able to self-administer the PedsQL™ in a clinic setting. In addition, internal consistency reliability was greater than 0.70 for most scales, supporting group comparisons. Scales with lower reliability (< 0.70) should be used for descriptive or exploratory analyses.

The Pain and Hurt Scale and the Pain Impact Scale demonstrated the strongest measurement properties for patient self-report. These two scales demonstrated no floor or ceiling effects, were internally consistent and differentiated between patients classified as having mild or severe SCD, supporting the sensitivity of these scales. Given that pain is a hallmark for sickle cell disease, these findings are very important and these scales will be critical in the evaluation of the HRQOL of children with SCD. The SCD Module Total Scale Score similarly demonstrated strong measurement properties for patients' self-report. For parent proxy-report, most of the scales demonstrated strong measurement properties, with only the Communication Scale I not differentiating between mild or severe disease.

Pediatric patients with SCD and their parents showed fair to moderate agreement across the scale scores for the PedsQL™ SCD Module Scales, with the strongest agreement for the Pain and Hurt Scale and the Pain Impact Scale. Given the salience of pain in this disease, and its observable, behavioral manifestations, it is perhaps not surprising that patient and parent agreement would be highest for this symptom domain. This finding is consistent with what we and others have found that suggests there is imperfect agreement between child self-reports and parent proxy-reports of children's HRQOL, especially for areas that are less observable and more internal (for example, physical functioning compared to emotional functioning)[54]. However, it is known that parents provide information that is complementary to the child's, but perhaps also unique [2]. Thus, we recommend that patient self-report be the primary measure of HRQOL and parent proxy-report a secondary measure.

The present study has several strengths, including the rigorous methods used to construct the measure, the large sample size, the broad age-range of participants, and the nationwide representation of participants from across the country. Limitations include lack of information on families who chose not to participate, and the small sample sizes for some of the subgroup analyses. Child self-report did not discriminate across all scales when comparing patients with mild and severe disease. It is well known in the field that there is no gold standard to determine disease severity in sickle cell disease and the method used here, consistent with prior work, relies on patients utilizing acute care services for episodes of pain. However, patients with sickle cell disease do experience pain at home for which they do not seek care and those patients in this study would have been classified as mild disease making it more difficult for our classification to differentiate between patients. Further work to determine the ability of the Module to differentiate across varied disease severity is needed, as well as research to determine the impact of disease modifying therapy on SCD-specific HRQOL. Parent proxy-report also showed more ceiling effects than the child self-report which may limit the Module's ability to detect greater improvement in health for

those particular scales. The ceiling effects observed were manifested in the scales with the fewest items (e.g., 2 or 3 items). However, in pediatric chronic health conditions, it is typically floor effects (when the lower values mean more of the symptom) that are of most concern. Some ceiling effects, but not floor effects, have been reported using the Generic Core Scales and Multidimensional Fatigue Scale in SCD [18]. Also, the intercorrelations among the SCD scales and the Generic Core and Fatigue scales may reflect, in part, shared method variance. In addition, our sample was drawn from a group of tertiary health care centers at arbitrary sites in the United States. This population may not represent the general population of patients and thus our findings may not be generalizable to all patients with SCD. However, we have no reason to believe the results are not generalizable. It will take widespread adoption of the module both in the United States and internationally to determine if HRQOL differs between populations. Lastly, more research to demonstrate additional psychometric properties of this Module are needed including test-retest reliability and responsiveness (ability to detect change over time) to further demonstrate the utility of the instrument. Ultimately, further work to determine the performance of this SCD-specific HRQOL instrument should help advance the concept that these questionnaires can be used in everyday clinical practice to monitor the HRQOL of a patient and to examine the impact of disease modifying therapies[55].

In conclusion, the PedsQL™ SCD Module has demonstrated initial feasibility, reliability and validity in pediatric patients with SCD. The PedsQL™ SCD Module Scales may be utilized in the evaluation of pediatric SCD disease-specific HRQOL in clinical research and practice to determine a patient's HRQOL. In conjunction with the PedsQL™ Generic Core Scales and the PedsQL™ Multidimensional Fatigue Scale, the new PedsQL™ SCD Module will facilitate the understanding of the health and well-being of children with SCD.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Abbreviations

PedsQL™	Pediatric Quality of Life Inventory™
SCD	Sickle Cell Disease
HRQOL	health-related quality of life

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Table I

PedsQL™ Sickle Cell Disease Module Scores, Reliability and Percent Floor and Ceiling Effects for Child Self-Report and Parent Proxy-Report

Sickle Cell Disease Module Scales	Number of Items	n	α	Mean	SD	% Floor	% Ceiling
Child Self-Report							
SCD Total Score	43	240	0.95	62.4	18.6	0	0.6
Pain and Hurt	9	243	0.86	66.7	20.9	0	5.3
Pain Impact	10	239	0.90	54.0	24.8	0.3	5.6
Pain Management and Control	2	235	0.78	54.9	29.9	7.5	10.6
Worry I	5	240	0.82	63.5	26.2	0.9	10.0
Worry II	2	182*	0.76	73.4	29.7	3.7	22.1
Emotions	2	238	0.69	62.0	33.1	8.4	19.3
Treatment	7	237	0.74	64.3	21.9	0.3	4.0
Communication I	3	239	0.70	73.8	24.9	0.3	23.4
Communication II	3	236	0.70	57.2	30.5	4.7	13.4
Parent Proxy-Report							
SCD Total Score	43	308	0.97	64.2	22.3	0	5.0
Pain and Hurt	9	306	0.94	67.7	23.6	0	12.8
Pain Impact	10	308	0.97	55.4	29.9	1.9	13.4
Pain Management and Control	2	308	0.93	61.3	31.7	7.2	27.7
Worry I	5	308	0.94	60.2	31.7	3.7	20.2
Worry II	2	307	0.89	69.3	33.1	9.0	38.3
Emotions	2	306	0.84	64.7	32.7	9.0	29.9
Treatment	7	308	0.87	69.0	23.2	0.3	15.3
Communication I	3	307	0.83	76.8	25.0	1.9	34.9
Communication II	3	307	0.83	65.8	30.2	3.7	28.0

Note: α = Cronbach's alpha internal consistency reliability. SD = standard deviation. ICC = intraclass correlations. Higher scores equal better HRQOL.

*For Worry II Scale Score child self-report, the sample size was smaller because this scale was available only for ages 8-18 years.

Table II

PedsQL™ Sickle Cell Disease Scales Scores Comparisons with Pediatric Patients with Mild and Severe Sickle Cell Disease for Child Self-Report and Parent Proxy-Report.

Scale	Severe		Mild		Differences	Effect Size	p-value
	Mean	SD	Mean	SD			
PedsQL™ Sickle Cell Disease							
Child Self-Report	*(n = 106)*		*(n = 137)*				
SCD Total Score	59.5	19.9	64.7	17.4	5.2	0.28	0.032
Pain and Hurt	61.6	22.4	70.7	18.8	9.0	0.44	0.001
Pain Impact	50.2	25.8	56.9	23.7	6.7	0.27	0.038
Pain Management and Control	51.8	29.3	57.2	30.3	5.4	0.18	0.171
Worry I	60.6	29.2	65.6	23.5	5.1	0.19	0.150
Worry II	71.0	27.2	75.4	31.6	4.4	0.15	0.316
Emotions	56.7	33.0	66.1	32.7	9.4	0.29	0.030
Treatment	63.0	21.3	65.3	22.4	2.3	0.11	0.432
Communication I	75.6	25.3	72.5	24.6	3.1	0.12	0.344
Communication II	55.1	30.6	58.9	30.5	3.8	0.12	0.345
Parent Proxy-Report	*(n = 129)*		*(n = 179)*				
SCD Total Score	57.2	21.5	69.3	21.5	12.1	0.56	<0.001
Pain and Hurt	58.9	23.1	74.1	22.0	15.2	0.67	<0.001
Pain Impact	48.2	27.5	60.6	30.6	12.4	0.43	<0.001
Pain Management and Control	55.5	30.7	65.4	31.9	9.9	0.32	0.007
Worry I	53.2	31.2	65.2	31.2	9.9	0.32	0.001
Worry II	62.7	33.8	74.2	31.8	11.5	0.35	0.003
Emotions	53.9	34.3	72.5	29.3	18.6	0.58	<0.001
Treatment	62.4	24.0	73.8	21.5	11.4	0.50	<0.001
Communication I	74.9	24.7	78.1	25.2	3.2	0.13	0.279
Communication II	59.4	29.3	70.4	30.1	11.0	0.37	0.001

Note: p-values based on independent samples *t*-tests. Effect sizes are designated as small (.20), medium (.50), and large (.80). Higher scores equal better HRQOL.

Table III

Pearson's Product Moment Correlations among PedsQL™ 4.0 Generic Core Scales and Sickle Cell Disease Module Scales for Child Self-Report (Above Diagonal) and Parent Proxy-Report (Below Diagonal) for Sickle Cell Disease Sample

Scale	GCT	PH	PsyH	EF	SF	SchF	SCD	PH	PI	PMC	WI	WII	EM	TR	COI	COII
Generic Core Total (GCT)	--	0.88	0.96	0.85	0.82	0.82	0.70	0.48	0.54	0.43	0.60	0.39	0.46	0.55	0.50	0.56
Physical Health (PhyH)	0.90	--	0.71	0.64	0.57	0.63	0.64	0.49	0.54	0.40	0.54	0.37	0.40	0.46	0.40	0.46
Psychosocial Health (PsyH)	0.96	0.74	--	0.88	0.87	0.84	0.65	0.43	0.48	0.40	0.57	0.36	0.44	0.54	0.50	0.55
Emotional Functioning (EF)	0.79	0.62	0.83	--	0.64	0.62	0.62	0.41	0.44	0.35	0.55	0.37	0.45	0.54	0.44	0.51
Social Functioning (SF)	0.82	0.65	0.86	0.56	--	0.58	0.47	0.26	0.32	0.31	0.45	0.26	0.30	0.37	0.46	0.46
School Functioning (SchF)	0.79	0.59	0.83	0.52	0.57	--	0.61	0.46	0.49	0.38	0.47	0.30	0.38	0.48	0.40	0.48
SCD Total Score (SCD)	0.68	0.61	0.65	0.64	0.46	0.56	--	0.78	0.88	0.67	0.80	0.53	0.63	0.71	0.54	0.67
Pain and Hurt (PH)	0.60	0.57	0.56	0.55	0.39	0.49	0.86	--	0.69	0.48	0.52	0.29	0.43	0.40	0.29	0.37
Pain Impact (PI)	0.58	0.53	0.55	0.52	0.39	0.49	0.90	0.76	--	0.58	0.62	0.38	0.49	0.48	0.36	0.48
Pain Management and Control (PMC)	0.52	0.47	0.50	0.48	0.33	0.46	0.81	0.65	0.76	--	0.43	0.28	0.42	0.46	0.34	0.39
Worry I (WI)	0.50	0.44	0.48	0.50	0.33	0.42	0.81	0.60	0.63	0.62	--	0.44	0.48	0.58	0.36	0.55
Worry II (WII)	0.43	0.39	0.42	0.44	0.25	0.37	0.64	0.46	0.44	0.45	0.76	--	0.27	0.41	0.34	0.33
Emotions (EM)	0.59	0.50	0.59	0.59	0.43	0.46	0.77	0.65	0.63	0.58	0.59	0.48	--	0.41	0.30	0.45
Treatment (TR)	0.62	0.55	0.59	0.59	0.43	0.50	0.83	0.66	0.65	0.68	0.63	0.51	0.67	--	0.36	0.46
Communication I (COI)	0.26	0.22	0.26	0.23	0.22	0.25	0.42	0.22	0.29	0.25	0.25	0.15	0.26	0.34	--	0.45
Communication II (COII)	0.51	0.43	0.51	0.53	0.38	0.39	0.70	0.47	0.54	0.51	0.51	0.39	0.54	0.59	0.56	--

Note: All values are statistically significant at $p<0.01$. Pearson's product moment correlations for child self-report are presented above the diagonal. Pearson's product moment correlations for parent proxy-report are presented below diagonal. Pearson's product moment correlation effect sizes are designated as small (0.10), medium (0.30), and large (0.50).

Table IV

Pearson's Product Moment Correlations among PedsQL™ Multidimensional Fatigue Scale and Sickle Cell Disease Module Scales for Child Self-Report (Above Diagonal) and Parent Proxy-Report (Below Diagonal) for Sickle Cell Disease Sample

Scale	TF	GF	SRF	CF	SCD	PH	PI	PMC	WI	WII	EM	TR	COI	COII
Total Fatigue Score (TF)	--	0.88	0.82	0.83	0.65	0.45	0.52	0.42	0.55	0.34	0.40	0.56	0.47	0.47
General Fatigue (GF)	0.90	--	0.63	0.61	0.70	0.50	0.58	0.44	0.60	0.34	0.45	0.56	0.43	0.53
Sleep/Rest Fatigue (SRF)	0.85	0.78	--	0.48	0.44	0.31	0.37	0.27	0.35	0.20	0.25	0.39	0.33	0.25
Cognitive Fatigue (CF)	0.82	0.57	0.45	--	0.52	0.33	0.36	0.36	0.44	0.33	0.31	0.47	0.44	0.42
SCD Total Score (SCD)	0.68	0.75	0.62	0.40	--	0.78	0.88	0.67	0.80	0.53	0.63	0.71	0.54	0.67
Pain and Hurt (PH)	0.62	0.70	0.59	0.34	0.86	--	0.69	0.48	0.52	0.29	0.43	0.40	0.29	0.37
Pain Impact (PI)	0.58	0.65	0.52	0.34	0.90	0.76	--	0.58	0.62	0.38	0.49	0.48	0.36	0.48
Pain Management and Control (PMC)	0.50	0.56	0.45	0.29	0.81	0.65	0.76	--	0.43	0.28	0.42	0.46	0.34	0.39
Worry I (WI)	0.48	0.54	0.46	0.26	0.81	0.60	0.63	0.62	--	0.44	0.48	0.58	0.36	0.55
Worry II (WII)	0.44	0.44	0.39	0.29	0.64	0.46	0.44	0.45	0.76	--	0.27	0.41	0.34	0.33
Emotions (EM)	0.56	0.61	0.48	0.36	0.77	0.65	0.63	0.58	0.59	0.48	--	0.41	0.30	0.45
Treatment (TR)	0.60	0.65	0.55	0.37	0.83	0.66	0.65	0.68	0.62	0.51	0.67	--	0.36	0.46
Communication I (COI)	0.34	0.32	0.28	0.28	0.42	0.22	0.29	0.25	0.25	0.15	0.26	0.34	--	0.45
Communication II (COII)	0.53	0.56	0.49	0.34	0.70	0.47	0.54	0.51	0.51	0.39	0.54	0.59	0.56	--

Note: All values are statistically significant at $p<0.01$. Pearson's product moment correlations for child self-report are presented above the diagonal. Pearson's product moment correlations for parent proxy-report are presented below diagonal. Pearson's product moment correlation effect sizes are designated as small (0.10), medium (0.30), and large (0.50).

Table V

Intraclass Correlations (ICCs) between Child Self-Report and Parent Proxy-Report on the PedsQL™ Sickle Cell Disease Module Scales for Pediatric Patients with Sickle Cell Disease

Sickle Cell Disease Module Scales	Parent-Child Agreement *ICCs
SCD Total Score	0.564
Pain and Hurt	0.596
Pain Impact	0.466
Pain Management and Control	0.382
Worry I	0.341
Worry II	0.348
Emotions	0.406
Treatment	0.431
Communication I	0.267
Communication II	0.275

* ICCs = Intraclass Correlation Coefficients. ICCs are designated as ≤0.40 poor to fair agreement, 0.41–0.60 moderate agreement, 0.61–0.80 good agreement, and 0.81–1.00 excellent agreement.