

# A Longitudinal Examination of Health-Related Quality of Life in Children and Adolescents With Spina Bifida

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**Objective** The current study examined (1) spina bifida (SB) youths' health-related quality of life (HRQOL) compared with nonclinical and chronic health condition (CHC) samples, (2) parent-child agreement regarding HRQOL, and (3) prospective changes in HRQOL. **Methods** Child and parent-proxy reports of Pediatric Quality of Life were collected at two time waves (Time 1:  $N = 134$ , ages 8–15 years; Time 2:  $N = 109$ , ages 10–17 years) as part of a larger longitudinal study. **Results** SB youth had statistically and clinically reduced physical HRQOL compared with the nonclinical and CHC samples at both time points. There were significant discrepancies between youth and parent-proxy reports of HRQOL; youth reported higher levels of physical and social HRQOL than parents. The majority of parent- and child-reported HRQOL domains remained stable, yet youth-reported social HRQOL increased over time. **Conclusions** Youth with SB are at risk for poor HRQOL. Examining modifiable condition and social-environmental predictors of youth HRQOL will be important in informing future interventions.

**Key words** longitudinal research; quality of life; spina bifida.

Spina bifida (SB) is a congenital birth defect that occurs in 3 of 10,000 live births in the United States (Centers for Disease Control and Prevention, 2011). This condition emerges during the first month of pregnancy when the neural tube fails to close completely (Burmeister et al., 2005) and is associated with motor, orthopedic, sensory, and cognitive impairments that impact youths' functioning (Fletcher & Brei, 2010). Myelomeningocele (MM) is the most common and severe type of SB; it is characterized by brain abnormalities and hydrocephalus. The majority of individuals with MM also have a distinct neurocognitive profile (e.g., executive function deficits) that may negatively impact social adjustment (Devine, Holmbeck, Gayes, & Purnell, 2012; Rose & Holmbeck, 2007). Thus, SB is a chronic condition associated with a multitude of physical, neurocognitive, and social challenges (Alriksson-Schmidt,

Wallander, & Biasini, 2007), and may create a significant burden on the well-being of these youth.

The experience of a chronic health condition (CHC) may have deleterious consequences on various aspects of a child's life, including health-related quality of life (HRQOL). HRQOL is characterized by several dimensions of a child's health and well-being (e.g., physical, psychological, and social well-being; De Civita et al., 2005) and has been recognized as a key marker of health outcomes in pediatric populations (Eiser & Jenney, 2007). Further, due to medical and technological advances, an increasing number of individuals with chronic illnesses are living longer, thus necessitating a focus on HRQOL as a salient outcome of interest. Advances in postnatal neurosurgical management have substantially reduced mortality among individuals with SB; the majority of children with this

condition are expected to live into adulthood (Bowman & McLone, 2010; Bowman, McLone, Grant, Tomita, & Ito, 2001). Thus, research should not only focus on the medical care of these individuals, but also on the development and enhancement of their quality of life.

Despite growing awareness of the importance of HRQOL assessment for several pediatric populations, much of the research to date has focused on only a few illness conditions (e.g., cancer, diabetes; Andelman, Zima, & Rosenblatt, 1999; Varni, Limbers, & Burwinkle, 2007a). For individuals with SB, the number of studies examining HRQOL has increased in the past decade. A recent literature search conducted by Sawin and Bellin (2010) found 39 studies addressing quality of life in individuals with SB of all ages, and about half of these studies focused on children and adolescents with this condition. A key finding from this review indicated that the majority of research in this area investigated demographic (e.g., age, gender, socioeconomic status [SES]; Kulkarni, Cochrane, McNeely, & Shams, 2008; Lemelle, Guillemin, Aubert, Guys, & Lottmann 2006; Sawin, Brei, Buran, & Fastnenau, 2002; Verhoef et al., 2007) and physical or medical correlates of decreased HRQOL (e.g., number of operations, bladder incontinence, mobility impairment, pain; Danielsson et al., 2008; Kirpalani et al., 2000; Oddson, Clancy, & McGrath, 2006; Verhoef et al., 2007). In contrast, few studies have aimed to compare HRQOL in children and adolescents with SB to nonclinical samples (i.e., typically developing individuals without chronic illnesses) or youth with CHCs using both *statistical* (i.e., *p* values) and *clinical* (i.e., minimal clinically important difference scores [MCIDs]; Varni, Burwinkle, Seid, & Skarr, 2003) indices of significance. Studies using nonclinical comparison samples have found that parents rated their children with SB as having statistically significantly lower HRQOL (Bartonek, Saraste, & Danielsson, 2012; Parekh et al., 2006). Further, children with SB have reported statistically lower HRQOL compared with youth with cerebral palsy (Okurowska-Zawada et al., 2011) and a mixed sample of youth with chronic illnesses (Parekh et al., 2006). Thus, it may be that youth with SB display statistically and clinically impaired HRQOL compared with nonclinical youth but only statistical impairments in HRQOL compared with chronically ill youth.

Further, because there may be minimal to moderate agreement between child and parent-proxy reports (Eiser & Morse, 2001), many researchers have emphasized the importance of collecting data on HRQOL from both the child and parent (Eiser & Varni, 2013). For youth with SB, only two studies to date have assessed parent-child agreement (Freeman et al., 2013; Parekh et al., 2006), indicating

moderate agreement. Thus, the second aim of this study was to provide data on parent-child agreement in children and adolescents with SB. Similar to the results of studies of youth with SB as well as other chronic illness groups (e.g., sickle cell disease; Panepinto, O'Mahar, DeBaun, Loberiza, & Scott, 2005), moderate agreement between child self-report and parent-proxy report of HRQOL was expected. Researchers have argued that agreement may depend on the clinical relevance of the domain. For example, physical functioning is highly salient for youth with rheumatoid arthritis, which may explain why parent-child agreement for this domain was high in one study (Varni et al., 2002). Because previous research has indicated that youth with SB are at risk for poor physical and social functioning (Bellin et al., 2010; Holmbeck et al., 2003), we expected that parent-child agreement for the physical and social HRQOL domains would be the highest compared with other domains (i.e., emotional, school). In addition, because a recent meta-analytic review of HRQOL in clinical and nonclinical pediatric populations indicated that parents of chronically ill children tend to report lower HRQOL scores than children themselves (Upton, Lawford, & Eiser, 2008), similar findings were expected for the current study.

Finally, to date, longitudinal studies of HRQOL in youth with SB have been limited to pre- and postoperative assessments of children undergoing urinary tract reconstruction (MacNeily, Scott, Dalgetty, & Afshar, 2009; Parekh et al., 2006). Thus, the final aim of this study was to examine prospective changes in HRQOL over a 2-year period (from Time 1 [ages 8–15 years] to Time 2 [ages 10–17 years]). There is a paucity of research on any pediatric condition that has examined the developmental course of HRQOL. Studies conducted thus far have yielded variable findings, likely because each pediatric condition is characterized by a unique clinical profile impacting long-term health outcomes. For example, longitudinal studies on youth with epilepsy have found that HRQOL remains stable over time (Modi, Ingerski, Rausch, & Glauser, 2011). Similarly, Sawyer and colleagues (2000) investigated a mixed illness sample of children and adolescents and found that there were no significant changes in physical HRQOL in children with asthma and diabetes over a 2-year period. On the other hand, children with cystic fibrosis reported that their physical health declined over time. Although SB is associated with several neurocognitive, physical, and social difficulties, researchers have noted that these youth may be better able to adapt to and accept their disease compared with other illness populations (Holmbeck, Coakley, Hommeyer, Shapera, & Westhoven, 2002). Therefore, the majority of HRQOL domains (i.e., social, emotional, and school-related) were

expected to remain stable in this population over a 2-year period. However, similar to research conducted on children with cystic fibrosis, physical HRQOL was expected to decline because youth with this condition may experience deterioration in physical health (i.e., due to an increased number of shunt revisions, tethered cord, or other surgeries).

Importantly, while knowledge concerning HRQOL in youth with SB has expanded, previous studies have significant methodological limitations. Limitations to the design and implementation of research in this area include the use of single informants, insufficient sample sizes (<30 participants), mixed samples (e.g., a mixed group consisting of youth with cerebral palsy or SB), the lack of clearly defined comparison samples, utilization of HRQOL measures that are not “well-established” (e.g., they tend not to include measures with good psychometric properties; Palermo et al., 2008; Sawin & Bellin, 2010), and cross-sectional study designs. The current study provides a methodologically rigorous longitudinal assessment of HRQOL in children and adolescents with SB (ages 8–15 years at Time 1; ages 10–17 years at Time 2). Methodological limitations found in previous research were addressed through the use of a longitudinal design, multiple informants (i.e., youth and parent report), a relatively large, illness-specific sample of youth with SB, nonclinical and CHC comparison samples, and a well-established measure of HRQOL (i.e., the Pediatric Quality of Life [PedsQL]).

## Method

### Participants

#### SB Sample

Participants in the SB sample were recruited starting in 2006 for a larger National Institutes of Health-funded study examining neurocognitive, family, and social domains in youth with SB (see Devine et al., 2012; Table 1). Participants were recruited from children’s hospitals and a statewide SB association in the Midwest using recruitment letters. Families were also approached and given information about the study during scheduled clinic visits. Interested participants were screened by phone or in person by a member of the research team, and were invited to participate if they met the following criteria: (1) diagnosis of SB, including MM, lipomeningocele, and myelocystocele, (2) ages 8–15 years, (3) involvement of at least one caregiver, (4) cognitive ability to complete questionnaires, (5) residence within 300 miles of the laboratory to allow for home visits, and (6) proficiency in English or Spanish. Eligible families were asked whether the child had reading comprehension difficulties and

Table 1. Demographic Characteristics: Mean (SD) or Percentage

Characteristic	SB sample <i>n</i> = 134	CHC comparison sample <i>n</i> = 214	Nonclinical comparison sample <i>n</i> = 684
Age (years)	11.2 (2.47) <sup>a</sup>	11.6 (2.38)	11.1 (2.49)
Gender			
Male (%)	44.8	49.1	43.4
Female (%)	55.2	50.9	56.6
Ethnicity			
White (%)	55.2	51.4	56.7
Hispanic (%)	27.6	30.0	27.5
Black (%)	14.2	14.0	13.7
Other/not reported (%)	3.0	5.6	2.1

Note. <sup>a</sup>Age of SB sample is based on Time 1 data when youth were 8–15 years old. CHC = chronic health condition.

preferred questionnaires to be read aloud by a research assistant.

Of the 246 families that were approached, 42% (*N* = 104) could not be contacted or declined participation. It was also determined that two of the families that participated did not meet inclusionary criteria. This resulted in an initial sample size of 140 families. Children of families that declined to participate did not differ from participants with respect to type of SB (MM or other),  $\chi^2(1) = 0.0002$ ,  $p > .05$ , shunt status,  $\chi^2(1) = 0.003$ ,  $p > .05$ , or occurrence of shunt infections,  $\chi^2(1) = 1.08$ ,  $p > .05$ . Other demographic and clinical variables, such as SES, race/ethnicity, and medical factors (e.g., ambulation status), were not collected from nonparticipants. Of the 140 families, the PedsQL 4.0 Generic Core Scales was completed by at least one family member from each of 134 families at Time 1. Specifically, 125 children aged 8–15 years old (mean age = 11.2; 55.2% female; see Table 1) completed the PedsQL at Time 1. Of these 125 children, 89 (71.2%) had a parent proxy PedsQL completed by both parents, 25 (20.0%) had mother report only, 4 (3.2%) had father report only, and 7 (5.6%) had no parent report. Nine families had a parent report but not a child report. Of the 134 families at Time 1, the PedsQL was completed by at least one family member from each of 109 families at Time 2. Of the 108 children aged 10–17 years (mean age = 13.4; 54.1% female) who completed the PedsQL at Time 2, 70 (64.8%) had a parent proxy PedsQL completed by both parents, 30 (27.8%) had mother report only, 6 (5.6%) had father report only, and 2 (1.9%) had no parent report. One family had a parent report but not a child report at Time 2.

Of the 134 families, 84.3% ( $N=113$ ) of children had a diagnosis of MM, 5.2% lipomeningocele ( $N=7$ ), and 6.0% other ( $N=8$ ; 5.1% or  $N=6$  unsure/missing). The greatest proportion of children had lumbar spinal lesions (48.6%,  $N=68$ ), whereas 29.3% ( $N=41$ ) and 16.4% ( $N=23$ ) had sacral and thoracic lesions, respectively (5.7% or  $N=8$  missing). Additionally, 77.6% ( $N=104$ ) of children had a shunt (1.4% or  $N=1$  missing). With respect to ambulation methods, 71.6% of the children used braces (i.e., ankle-foot, knee-ankle-foot, and/or hip-knee-ankle foot orthotics), 44.8% used a walker or crutches, and 56.7% used a wheelchair (ambulation categories were nonexclusive). Finally, the mean Hollingshead SES (Hollingshead, 1975) score for the sample was 40.0 (i.e., upper middle class status) at Time 1. Hollingshead SES could not be computed for 5.8% of the sample ( $N=8$ ) because of missing data on parents' occupation and/or education.

### Nonclinical and CHC Comparison Samples

To provide matched comparison samples for the sample of youth with SB, we used previously published data for 684 nonclinical youth with no CHCs (mean age = 11.1; 56.6% female) and 214 youth with CHCs (mean age = 11.6; 50.9% female; Varni et al., 2003; Table 1). Youth from the nonclinical sample did not receive care from a specialty clinic providing services for children with acute or CHCs and were not diagnosed with an acute health condition or a CHC. The CHC sample included children with asthma, attention-deficit/hyperactivity disorder, depression, diabetes, or other CHCs (Varni et al., 2003). The PedsQL was completed by parents and children via the mail. Please see Varni and colleagues (2003) for further details regarding recruitment and data collection procedures. We used data from the PedsQL database that were randomly matched to our sample of youth with SB on age, gender, and ethnicity. Participants were matched at a ratio in which the number of individuals from the nonclinical and CHC samples was greater than the sample size of youth with SB. The appropriate institutional review committees approved the original published study.

### Procedure

The sponsoring institution and hospitals' institutional review boards approved this study. Trained graduate and undergraduate research assistants conducted home visits at each data collection wave. Caregiver informed consent, child assent, and medical release forms were obtained before data collection at each visit. Mothers, fathers, and youth completed a questionnaire battery to gain information on several areas of psychosocial functioning. The

current study is based on parent and youth report on a measure of HRQOL (i.e., the PedsQL) that was included in questionnaire packets. Parents also completed a demographics and medical form.

### Health-Related Quality of Life

Youths' HRQOL was assessed using youth, mother, and father report on the PedsQL Scale (PedsQL™ 4.0 Generic Core Scales; Varni, Seid, & Kurtin, 2001). The PedsQL has well-established reliability and validity in children with both acute health conditions and CHCs, and yields an 8-item physical scale as well as a 15-item psychosocial scale. The psychosocial scale is further composed of three subscales: emotional (five items), social (five items), and school functioning (five items). Youth and parents answered how much of a problem a given task had been over the past month using a 5-point Likert scale rating (0 = *never a problem* to 4 = *almost always a problem*). The following are sample questions from each scale of the child version: "I hurt or ache" (physical); "I feel sad or blue" (emotional); "Other kids do not want to be my friend" (social); "I have trouble keeping up with my schoolwork" (school). Raw scores were transformed into standard scores that ranged from 0 to 100, with higher scores indicating better HRQOL. Child (ages 8–12 years) or adolescent (ages 13–18 years) versions of the PedsQL were administered according to age. In the current study, internal consistency was adequate for each scale ( $\alpha$  values = .83–.90) across reporters and time points. The Flesch-Kincaid reading level of the PedsQL ranges from below-first to mid-second grade levels for the child version and third to sixth grade for the adolescent version (Varni, Seid, & Rode, 1999).

### Statistical Treatment

Statistical analyses were conducted using SPSS Version 21.0. Before hypothesis testing, preliminary analyses determined whether HRQOL scores were skewed or contained outliers. Independent  $t$  tests were used to assess *statistically* significant group differences. *Clinically* significant impairment in HRQOL was assessed using MCIDs available for child and parent proxy-report versions of the PedsQL (Varni et al., 2003). MCIDs represent the amount of change in HRQOL needed to initiate care seeking or a change in care to improve functioning (Varni et al., 2003). Effect sizes were calculated by taking the difference between the reference (i.e., nonclinical or CHC) sample mean and the SB sample mean, divided by the standard deviation of the reference sample mean. Effect sizes for differences in means were classified as small (0.20),

medium (0.50), and large (0.80) in magnitude (Cohen, 1988).

Degree of agreement between parent and child ratings of HRQOL was examined using intraclass correlation coefficients (ICCs) and paired sample *t* tests. The ICC provides an index of absolute agreement, as it takes into account the ratio between subject variability and total variability. Following established guidelines for kappa (Landis & Koch, 1977), ICCs between 0.00 and 0.20 indicated “slight” agreement, between 0.21 and 0.40 indicated “fair” agreement, between 0.41 and 0.60 indicated “moderate” agreement, between 0.61 and 0.80 indicated “substantial” agreement, and between 0.81 and 1.00 indicated “almost perfect” agreement (Landis & Koch, 1977). Finally, two repeated measures multivariate analyses of variance (MANOVAs) were conducted (i.e., two separate MANOVAs for youth and parent report) to determine whether youth and parent-proxy reports of physical, emotional, social, and school-related HRQOL remained stable or changed (i.e., significantly increased or decreased) from Time 1 (i.e., youth ages 8–15 years) to Time 2 (i.e., youth ages 10–17 years). MANOVAs contained two within-subjects factors: time and the above four HRQOL scales.

## Results

### Preliminary Analyses

Study results indicated that, across respondents, reports of youths’ HRQOL at Time 1 and Time 2 were not highly skewed (range of skewness values = .43 to  $-.58$ ). Results revealed substantial to almost perfect agreement between mother- and father-proxy report of HRQOL (ICCs = .66–.84;  $M = 0.74$ ) and were, therefore, averaged together to reduce the number of analyses conducted. Descriptive data by group are reported in Table I. Independent *t* tests and chi-squared analysis ( $3 \times 2$ ) were used to determine group differences in age, gender, and ethnicity. There were no significant differences between the sample of youth with SB and the nonclinical and CHC samples according to age [ $t(816) = -0.37, p > .05$  and  $t(346) = 1.65, p > .05$ , respectively], gender [ $\chi^2(2) = 2.10, p > .05$ ], or ethnicity [ $\chi^2(6) = 8.10, p > .05$ ] at Time 1.

In addition, there was a significant difference for Time 1 parent report (but not child report) of total psychosocial HRQOL between those who did not participate at Time 2 and those who participated at both time points; parent nonresponders reported lower levels of HRQOL at Time 1 compared with responders [ $N$  of nonresponders = 29;  $t(125) = -2.21, p = .029$ ]. Despite this statistical difference between parent responders and nonresponders, statistical analyses were conducted as planned.

### Aim 1: Group Differences in HRQOL: SB and Nonclinical Samples

#### Youth Report

As shown in Table II, youth with SB reported significantly lower HRQOL across all domains compared with the nonclinical sample at both time points ( $ps < .05$ ). In addition, between-group difference scores met the criterion for being considered a minimal clinically important difference for all HRQOL domains and at both time points. Effect sizes for group differences were medium to large (d-values = 0.69–2.85).

#### Parent Report

Similar to youth report, parents reported significantly lower HRQOL across all domains compared with the nonclinical sample at both time points ( $ps < .05$  Table III). Absolute difference scores met the criterion for being considered a minimal clinically important difference across HRQOL domains and time points. Effect sizes for group differences were also medium to large (d-values = 0.64–2.23).

### Aim 1: Group Differences in HRQOL: SB and CHC Samples

#### Youth Report

Youth with SB reported significantly lower HRQOL across all domains compared with the CHC sample at Time 1 and significantly lower physical and school-related HRQOL at Time 2 ( $ps < .05$  Table II). Between-group difference scores met the criterion for being considered a minimal clinically important difference for physical, psychosocial total, and social HRQOL at Time 1 and for physical HRQOL at Time 2. Effect sizes for group differences varied considerably from no effect to large (i.e., d-values = 0.00–1.28). The effect sizes were large for the group comparison of physical HRQOL at both time points (d-values = 1.28 and 1.13 at Time 1 and Time 2, respectively) and small for the group comparisons involving psychosocial total, and social HRQOL at Time 1 (d-values = 0.43 and 0.45, respectively).

#### Parent Report

As shown in Table III, parents reported significantly lower physical, psychosocial total, social, and school-related (i.e., all domains except emotional) HRQOL compared with the CHC sample at Time 1, and significantly lower physical and social HRQOL at Time 2 ( $ps < .05$ ). Between-group difference scores met the criterion for being considered a minimal clinically important difference for physical, psychosocial total, and social HRQOL at Time 1 (similar to child report) and for physical and social HRQOL at Time 2. Effect sizes for group differences varied considerably from no effect to large (d-values = 0.09–1.18; Table III). Effect

Table II. Unadjusted Group Means for Child Report on the PedsQL 4.0 Genetic Core Scales

HRQOL domain	Time 1					Time 2				
	Mean (SD)	MCID <sup>a</sup>	Absolute difference (vs. SB)	<i>d</i>	<i>p</i>	Mean (SD)	MCID <sup>a</sup>	Absolute difference (vs. SB)	<i>d</i>	<i>p</i>
Physical scale		6.66					6.66			
SB sample	58.61 (21.49)					61.04 (20.64)				
CHC sample	79.50 (16.35)		20.89 <sup>b</sup>	1.28	.000	79.50 (16.35)		18.46 <sup>b</sup>	1.13	.000
Nonclinical sample	90.23 (11.08)		31.62 <sup>b</sup>	2.85	.000	90.23 (11.08)		29.19 <sup>b</sup>	2.63	.000
Psychosocial total scale		5.30					5.30			
SB sample	62.77 (16.73)					67.61 (16.17)				
CHC sample	70.49 (17.16)		7.71 <sup>b</sup>	0.45	.000	70.49 (17.16)		2.87	0.17	.158
Nonclinical sample	83.32 (12.79)		20.55 <sup>b</sup>	1.61	.000	83.32 (12.79)		15.71 <sup>b</sup>	1.23	.000
Emotional subscale		8.94					8.94			
SB sample	64.40 (20.47)					69.03 (19.28)				
CHC sample	69.11 (19.74)		4.72	0.24	.043	69.11 (19.74)		0.08	0.00	.972
Nonclinical sample	80.83 (17.19)		16.43 <sup>b</sup>	0.96	.000	80.83 (17.19)		11.80 <sup>b</sup>	0.69	.000
Social subscale		8.36					8.36			
SB sample	66.45 (22.40)					72.52 (19.82)				
CHC sample	76.02 (22.13)		9.57 <sup>b</sup>	0.43	.000	76.02 (22.13)		3.50	0.16	.175
Nonclinical sample	87.41 (14.89)		20.96 <sup>b</sup>	1.41	.000	87.41 (14.89)		14.89 <sup>b</sup>	1.00	.000
School subscale		9.12					9.12			
SB sample	57.56 (21.80)					61.31 (20.72)				
CHC sample	66.34 (19.62)		8.78	0.45	.000	66.34 (19.62)		5.03	0.26	.039
Nonclinical sample	81.74 (14.87)		24.18 <sup>b</sup>	1.63	.000	81.74 (14.87)		20.43 <sup>b</sup>	1.37	.000

Notes. <sup>a</sup>MCIDS = minimal clinically important difference scores (Varni, Burwinkle, Seid, & Skarr, 2003).

<sup>b</sup>Denotes that between-group difference score met minimum criterion for being considered a meaningful clinically important difference.

*d* = effect sizes. CHC = chronic health condition. Higher values indicate better health-related quality of life. *N*s = 124–125 for the SB sample at Time 1 and 107–108 at Time 2; *N*s = 189–190 for the CHC sample; *N* = 662 for the nonclinical sample. Effect sizes are designated as small (.20), medium (.50), and large (.80; Cohen, 1988).

sizes were large for the group comparison of physical HRQOL at both time points (*d*-values = 1.15 and 1.18 at Times 1 and 2, respectively) and social HRQOL at Time 1 (*d* = 0.59). Effect sizes were small for the group comparison of psychosocial total HRQOL at Time 1 (*d* = 0.39) and social HRQOL at Time 2 (*d* = 0.42).

### Aim 2: Parent–Child Agreement on HRQOL for the SB Sample

*Intraclass Correlations (ICCs) Between Parent and Child Ratings of HRQOL.* Results indicated that there was slight to moderate agreement between parent-proxy and child report across all domains of HRQOL at Time 1 (ICC = 0.20–0.54; see Table IV). Specifically, at Time 1, ICCs for physical and school functioning indicated moderate agreement. Agreement was fair for psychosocial total and emotional functioning and slight for social functioning. Results also indicated there was moderate interrater agreement for all HRQOL domains at Time 2.

*Discrepancies between parent and child reports of HRQOL.* Analyses of mean differences between child and parent reports of HRQOL indicated that there were no significant differences between youth and parent reports for Time 1 psychosocial total [ $t(120) = 1.21, p > .05$ ], emotional [ $t(120) = -0.65, p > .05$ ], and school functioning [ $t(120) = -0.27, p > .05$ ]. In addition, there were no significant differences for Time 2 emotional [ $t(102) = -0.71, p > .05$ ] and school functioning [ $t(102) = 0.66, p > .05$ ; See Table IV].

However, there were significant differences between parent and youth reports for Time 1 and Time 2 physical HRQOL [ $t(120) = 2.68, p < .01$ ;  $t(102) = 4.07, p < .05$ , respectively], Time 2 psychosocial total HRQOL, [ $t(102) = 2.15, p < .05$ ], and Time 1 and Time 2 social HRQOL [ $t(120) = 3.64, p < .01$ ;  $t(102) = 4.88, p < .01$ , respectively]. Child report of HRQOL, and, in particular, physical and social functioning, was significantly higher than parent report at both time points (i.e., youth reported

Table III. Unadjusted Group Means for Parent Proxy Report on the PedsQL 4.0 Genetic Core Scales

HRQOL domain	Time 1					Time 2				
	Mean (SD)	MCIDS <sup>a</sup>	Absolute difference (vs. SB)	<i>d</i>	<i>p</i>	Mean (SD)	MCIDS <sup>a</sup>	Absolute difference (vs. SB)	<i>d</i>	<i>P</i>
Physical scale		6.92					6.92			
SB sample	52.71 (20.46)					52.25 (20.59)				
CHC sample	76.07 (20.26)		23.36 <sup>b</sup>	1.15	.000	76.07 (20.26)		23.82 <sup>b</sup>	1.18	.000
Nonclinical sample	87.69 (15.88)		34.98 <sup>b</sup>	2.20	.000	87.69 (15.88)		35.44 <sup>b</sup>	2.23	.000
Psychosocial total scale		5.49					5.49			
SB sample	60.90 (12.72)					64.51 (13.71)				
CHC sample	67.77 (17.56)		6.86 <sup>b</sup>	0.39	.000	67.77 (17.56)		3.25	0.19	.071
Nonclinical sample	81.70 (13.84)		20.79 <sup>b</sup>	1.50	.000	81.70 (13.84)		17.18 <sup>b</sup>	1.24	.000
Emotional subscale		7.79					7.79			
SB sample	66.21 (14.91)					70.62 (15.30)				
CHC sample	68.02 (20.15)		1.82	0.09	.378	68.02 (20.15)		2.60	0.13	.201
Nonclinical sample	80.94 (16.15)		14.74 <sup>b</sup>	0.91	.000	80.94 (16.15)		10.32 <sup>b</sup>	0.64	.000
Social subscale		8.98					8.98			
SB sample	58.67 (15.71)					62.66 (18.88)				
CHC sample	72.11 (22.61)		13.44 <sup>b</sup>	0.59	.000	72.11 (22.61)		9.45 <sup>b</sup>	0.42	.000
Nonclinical sample	85.56 (16.83)		26.89 <sup>b</sup>	1.60	.000	85.56 (16.83)		22.90 <sup>b</sup>	1.36	.000
School subscale		9.67					9.67			
SB sample	57.68 (17.33)					59.97 (17.54)				
CHC sample	63.23 (20.87)		5.54	0.27	.009	63.23 (20.87)		3.25	0.16	.146
Nonclinical sample	78.58 (16.98)		20.90 <sup>b</sup>	1.23	.000	78.58 (16.98)		18.61 <sup>b</sup>	1.10	.000

Notes. <sup>a</sup>MCIDS = minimal clinically important difference scores (Varni, Burwinkle, Seid, & Skarr, 2003).

<sup>b</sup>Denotes that between-group difference score met minimum criterion for being considered a meaningful clinically important difference.

*d* = effect sizes. CHC = chronic health condition. Higher values equal better health-related quality of life. *N*s = 126–127 for the SB sample at Time 1 and 106–107 at Time 2; *N*s = 18–190 for the CHC sample; *N* = 662 for the nonclinical sample. Effect sizes are designated as small (.20), medium (.50), and large (.80; Cohen, 1988).

better functioning compared with parent-proxy). Children also reported higher levels of psychosocial functioning at Time 2.

### Aim 3: The Course of HRQOL in Youth With SB Over a 2-Year Period

Repeated measures MANOVA was performed to address the hypothesis that HRQOL (i.e., physical, emotional, social, and school-related) would remain stable over time. MANOVAs included the within-subjects factors of time and HRQOL subscale. The repeated measures MANOVA using youth-reported HRQOL indicated a significant within-subject change over time [ $F(1, 95) = 4.62, p < .05$ ]. Subsequent follow-up univariate analyses of variance indicated that, contrary to hypotheses, there was a significant increase in social HRQOL [ $F(1, 97) = 4.38, p < .05$ ]. Youth-reported physical [ $F(1, 97) = 0.08, p > .05$ ], emotional [ $F(1, 96) = 3.90, p = .051$ ], and school-related [ $F(1, 96) = 3.89, p = .051$ ] HRQOL did

not significantly change from Time 1 to Time 2. The MANOVA within-subject effect for time was not significant for parent-proxy report of HRQOL [ $F(1, 97) = 0.85, p > .05$ ].

### Discussion

The present study adds to the growing body of literature on HRQOL in children and adolescents with SB, and addresses several methodological limitations of previous research. We aimed to examine HRQOL in this population utilizing longitudinal multi-informant data in a relatively large illness-specific sample of youth with SB. We also compared this sample with nonclinical and chronically ill (age, gender, and ethnicity) matched comparison samples with a well-established measure of HRQOL (i.e., the PedsQL). Consistent with previous research on other pediatric populations, such as cancer, sickle cell, obese, and chronic pain patients (Hunfeld et al., 2001; Modi &

Table IV. Summary of Parent–Child Agreement on HRQOL

HRQOL domain	Child mean scores (SD)	Caregiver mean scores (SD)	Interrater agreement	
			ICC	Paired-sample <i>t</i> test ( <i>t</i> value)
Physical scale				
Time 1	58.61 (21.49)	52.71 (20.46)	.48**	2.68*
Time 2	61.04 (20.64)	52.25 (20.59)	.51**	4.07**
Psychosocial total scale				
Time 1	62.77 (16.73)	60.90 (12.72)	.38*	1.21
Time 2	67.61 (16.17)	64.51 (13.71)	.59**	2.15*
Emotional subscale				
Time 1	64.40 (20.47)	66.21 (14.91)	.30*	−.65
Time 2	69.03 (19.28)	70.62 (15.30)	.44*	−.71
Social subscale				
Time 1	66.45 (22.40)	58.67 (15.71)	.20	3.64**
Time 2	72.52 (19.82)	62.66 (18.88)	.52**	4.88**
School subscale				
Time 1	57.56 (21.80)	57.68 (17.33)	.54**	−.27
Time 2	61.31 (20.72)	59.97 (17.54)	.60**	.66

Notes. ICCs between 0.00 and 0.20 indicated “slight” agreement, between 0.21 and 0.40 indicated “fair” agreement, between 0.41 and 0.60 indicated “moderate” agreement, between 0.61 and 0.80 indicated “substantial” agreement, and between 0.81 and 1.00 indicated “almost perfect” agreement (Landis & Koch, 1977). \* $p < .01$ ; \*\* $p < .001$ .

Quittner, 2003; Palermo, Schwartz, Drotar, & McGowan, 2002; Schwimmer, Burwinkle, & Varni, 2003; Varni, Limbers, & Burwinkle, 2007b), results indicated that children and adolescents with SB may be at risk for reduced HRQOL across the childhood and adolescent developmental periods. Supporting hypotheses, HRQOL scores across all domains were statistically and clinically lower compared with the nonclinical sample, and this finding was consistent across youth and parent-proxy reports at both time points. Contrary to hypotheses, child report and parent-proxy report of HRQOL were also clinically lower than youth with CHCs, particularly at a younger age (i.e., Time 1, when youth were aged 8–15 years) and within the physical and social domains of functioning. A focus on physical and social functioning will be important to include in interventions designed to improve the health and wellness of youth with SB. Clinicians should pay special consideration to improving HRQOL in younger children and adolescents with this condition, and future studies could examine whether the course of HRQOL varies according to developmental period (e.g., childhood, adolescence).

The degree to which parents and youth agree on reports of HRQOL was also examined. Taken together,

correlational and paired *t*-test analyses indicated that parent–child agreement was slight to moderate when youth were aged 8–15 years (i.e., Time 1) and moderate when participants were aged 10–17 years (i.e., Time 2). Closely related parent and child perceptions of a particular domain of functioning may be the result of higher levels of parental involvement around relevant issues of health, health care (Upton et al., 2008), and normative development. Specifically, agreement was moderate for physical and school-related functioning at both time points, which may highlight the clinical relevance of these domains for children and adolescents with SB across several periods of development. Parent–child agreement for emotional and social functioning was moderate at Time 2 but not at Time 1, possibly due to the increased relevance of developmentally normative difficulties with psychological functioning and peer relationships as youth progress through late childhood and into adolescence.

Relatedly, we also found significant mean-level discrepancies between youth and parent-proxy reports for many domains of HRQOL. Interestingly, past research on cross-informant discrepancies has been inconsistent. Some research has indicated that chronically ill children may report more physical and emotional distress compared with their parents (Modi & Quittner, 2003; Theunissen et al., 1998; Verrips, Vogels, den Ouden, Paneth, & Verloove-Vanhorick, 2000), whereas other studies have found that youth report better HRQOL compared with parent-proxy report (e.g., cerebral palsy, cystic fibrosis, and chronic pain; Berrin et al., 2007; Britto, Kotagal, Chenier, Tsevat, Atherto, & Wilmott, 2004; Gold, Mahrer, Yee, & Palermo, 2009; Sawyer et al., 2005). The current study found that parents reported significantly lower social and physical HRQOL compared with youth report at both periods. It is also possible that the child with SB adapts to his or her physical and social limitations, while parents continue to compare their child’s quality of life with that of typically developing siblings or peers. Furthermore, due to the neurocognitive impairments associated with SB, discrepancies between parent and youth reports may reflect impairments in higher-level metacognitive abilities needed for self-assessment (Lennon, Klages, Amaro, Murray, & Holmbeck, 2015). Perhaps more importantly, noted discrepancies highlight the need to follow the suggestion provided by many experts in the field (e.g., Modi & Quittner, 2003; Palermo et al., 2008) that data should be collected from both children and parents. Parent–child disagreement on HRQOL does not necessarily imply that one rater may be more accurate or “correct” in his/her perception, but rather that parents and

children may each contribute different but valid information (Eiser & Varni, 2013; Upton et al., 2008).

Partially supporting hypotheses, parent-proxy report of HRQOL remained stable over time. Yet contrary to hypotheses, child report of social HRQOL increased and physical HRQOL did not significantly change over time. Results may point to a resiliency factor in families of youth with SB as indicated in previous research (Holmbeck et al., 2002); youth with SB may be better able to adapt to and/or accept their disease compared with other illness populations. Still, it should be noted that select domains of HRQOL were significantly and clinically lower compared with the nonclinical and CHC samples at both time points. Therefore, although youth report of social HRQOL increased, youth with SB still had relatively low social functioning across time. The stability of parent-proxy reports of HRQOL in this study is consistent with findings from other longitudinal studies that have also found similar trends in pediatric illness groups (Sawyer et al., 2005). For example, a study of caregivers of 124 newly diagnosed epileptic children found that HRQOL remained stable over time (Modi et al., 2011). Finally, because some studies have suggested that individuals with SB may be at risk for emotional distress and poor HRQOL during emerging and young adulthood (Bellin, Bentley, & Sawin, 2009; Bellin et al., 2013), it will be important for research to investigate trajectories of HRQOL from childhood to adulthood. Future research should determine whether trajectories of HRQOL remain stable, increase, or have a bell-shaped curve due to the difficulties individuals might have with securing employment and gaining independence as well as the possible deterioration of their condition (Kaneko, Sato, Soejima, & Kamibepu, 2014).

This study represents an important step in documenting that youth with SB may be at risk for clinically low levels of HRQOL. However, several limitations of this study should be considered. First, parents who completed the protocol only at Time 1 reported lower levels of HRQOL compared with those who participated at both time points. Thus, the results of this study may not generalize to youth with particularly poor quality of life because these families may have dropped out of the study. On the other hand, the fact that our findings were still significant even with such selective attrition suggests that our findings may be particularly robust. Second, as researchers have previously noted, the PedsQL contains questions that may not be appropriate for those with physical disabilities (e.g., “It is hard for me to walk more than one block”; “It is hard for me to run”); the results involving the physical scale may have been inflated because the majority of the study sample had physical limitations due to their

condition (Oddson et al., 2006). Importantly, a SB disease-specific measure of HRQOL that parallels the PedsQL is still needed for this population. Third, HRQOL data were based on group means, yet individual trajectories may vary; the HRQOL of some patients may increase whereas others remain stable or decline over time. Finally, the age range of the current study is large and crosses several developmental stages. Future studies should investigate whether stability or change in HRQOL varies as a function of important demographic or disease-related characteristics (e.g., age, gender, type of SB, number of surgeries, and access to health care); Seid, Varni, Cummings, & Schonlau (2006).

The results of this study suggest that children and adolescents with SB are at risk for clinically reduced HRQOL, and it is possible that such trends will continue into adulthood. Determining factors that influence HRQOL will be an important next step in developing appropriate interventions that improve functioning for this population. The majority of research that has examined predictors of HRQOL in youth with SB has tended to focus on nonmodifiable demographic and illness-specific correlates, such as age, gender, and level of mobility impairment (see Bellin et al., 2010). Research examining the influence of modifiable condition and social–environmental predictors (e.g., pain, sleep, weight status, family environment) of youths’ HRQOL may be particularly relevant for future intervention development.

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