Qual Life Res
DOI 10.1007/s11136-015-0988-5

QUAlity of Life Assessment in Spina bifida for Adults (QUALAS-A): development and international validation of a novel health-related quality of life instrument

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Accepted: 7 April 2015

Abstract

Purpose Health-related quality of life (HRQOL) is important in spina bifida (SB) management. No clinically useful, comprehensive instrument incorporating bladder/bowel domains exists. We aimed to develop and validate a self-reported QUAlity of Life Assessment in Spina bifida for Adults (QUALAS-A).

Methods We drafted the 53-question pilot instrument using a comprehensive item generation/refinement process. It was administered to an international convenience sample of adults with SB and controls recruited online via social media and in person at outpatient SB clinics (January 2013–September 2014). Final questions were determined by: clinical relevance, high factor loadings and domain psychometrics in an Internal Validation Sample randomly selected from United States participants ($n = 250$). External validity was evaluated in United States and International External Validation Samples ($n = 165$ and $n = 117$, respectively). Adults with SB completed the validated general WHOQOL-BREF and International Consultation on Incontinence Questionnaire (ICIQ).

Results Mean age of 532 participants was 32 years (32.7 % males, 85.0 % Caucasian), similar to 116 controls ($p \geq 0.08$). There were 474 online and 58 clinical participants (61.1 % eligible). Face validity and content validity of the 3-domain, 15-question QUALAS-A were established by patients, families and experts. Internal consistency and test–retest reliability were high for all domains (Cronbach's alpha ≥ 0.70, ICC ≥ 0.77). Correlations between QUALAS-A and WHOQOL-BREF were low ($r \leq 0.60$), except for high correlations with Health and Relationships domain ($0.63 \leq r \leq 0.71$). Bladder and Bowel domain had a high correlation with ICIQ ($r = -0.70$). QUALAS-A scores were lower among adults with SB than without ($p < 0.0001$). QUALAS-A had good statistical properties in both External Validation Samples (Cronbach's alpha 0.68–0.77).

Conclusions QUALAS-A is a short, valid HRQOL tool for adults with SB.

Keywords Quality of life · Spinal dysraphism · Meningomyelocele · Adult · Urinary incontinence · Fecal incontinence

Abbreviations

HRQOL	Health-related quality of life
QOL	Quality of life

Electronic supplementary material The online version of this article (doi:10.1007/s11136-015-0988-5) contains supplementary material, which is available to authorized users.

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SB	Spina bifida
QUALAS-A	QUAlity of Life Assessment in Spina bifida for Adults
WHOQOL-BREF	World Health Organization Quality of Life instrument
SD	Standard deviation

Purpose

Spina bifida (SB) is the most common congenital anomaly of the central nervous system, affecting 3.4 per 10,000 children born in the United States [1]. Children who survive infancy face major challenges, including developmental delay, paresis, bowel and bladder dysfunction. As with other chronic conditions, one of the aims of successful management of SB is to improve health-related quality of life (HRQOL). However, this approach is based on little evidence. Studies tend to use non-validated HRQOL instruments [2] or simply report patient satisfaction [3, 4]. HRQOL literature on SB is mostly comprised of small, single-institutional studies using multiple, limited and poorly validated instruments [5]. In addition, generic HRQOL instruments developed for healthy adults [6, 7] lack sensitivity to measuring the impact of SB on HRQOL and thus may be unable to capture small, but clinically relevant, differences [8]. In fact, no comprehensive, validated and clinically useful SB-specific HRQOL instrument incorporating Bladder and Bowel domains exists.

Assessing an individual's own HRQOL is particularly important in patient-centered care, as those with SB often report better HRQOL than that perceived by their parents and caregivers [9, 10]. For this reason, instrument development requires input from multiple stakeholders, including those with SB, as well as their parents and caregivers, rather than only healthcare professionals [5, 8]. A suitable, clinically relevant HRQOL instrument focuses on HRQOL, rather than physical function, possesses excellent psychometric properties and is condition specific, while remaining short and straightforward [5, 8]. Our goal was to develop and validate a clinically useful, self-reported, disease-specific, health-related QUAlity of Life Assessment in Spina bifida for Adults (QUALAS-A).

Methods

The study received Indiana University Internal Review Board approval (IRB 9470) and followed the Federal Drug Administration recommendations for patient-reported outcome instrument development [11].

Inclusion and exclusion criteria

Adults (≥18 years old) with a history of myelomeniongocele or lipomeningocele requiring newborn spine surgery were included. Exclusion criteria were: poor self-reported English proficiency (rated as "a little bit" or "some," rather than "well" or "very well" on a 4-point Likert scale), developmental delay that interferes with answering questions, not completing the questionnaire, surgery in the last month or a primary diagnosis other than spina bifida (primary tethered cord, sacral agenesis, medullary lipoma, anorectal malformation, spinal trauma or tumor). Inclusion and exclusion criteria remained unchanged for all phases of this study. Healthy controls without a personal history of SB or a child with SB were recruited from hospital staff and colleagues.

Phase 1: Item generation

We used a patient-centered, comprehensive item generation and refinement process (Fig. 1). Items were generated from a series of 15 semi-structured interviews with adults with SB and their families (14 in clinic, 1 by telephone), and a focus group with two adults with SB and two parents. Items were also drafted based on a review of published instruments and the opinions of 20 national and international experts in SB care (see Acknowledgments). The item generation process continued until reaching the saturation point, when no new items were generated. This comprehensive list of 150 items covered themes of cognition, independence, emotional impact, social interactions, educational, work and leisure activities, mobility, future plans, sexuality, healthcare interactions, toileting as well as urinary and fecal continence. This exhaustive list was reduced to 89 items by experts, who eliminated similarly worded and redundant items, while maintaining the themes.

Phase 2: Development of pilot instrument

The 89 items were rated on importance by eight adults with SB and two parents from our center's SB clinic. Ratings were made on a 3-point Likert scale (not important, somewhat important, very important). After review by a panel of experts, 53 items with the highest rankings per theme were selected in order to create a more manageable and representative pilot QUALAS-A. This item reduction approach has been successfully used in the SB population [12–14]. Some items were reworded to ensure comprehension and clarity. Rewording was based on the feedback from the 10 adult raters, by an additional two adult pilot participants and a Health Literacy Educator at our institution's Family Education Center (see Acknowledgments). Readability was assessed by the Flesch Kincaid Grade Level test [15].

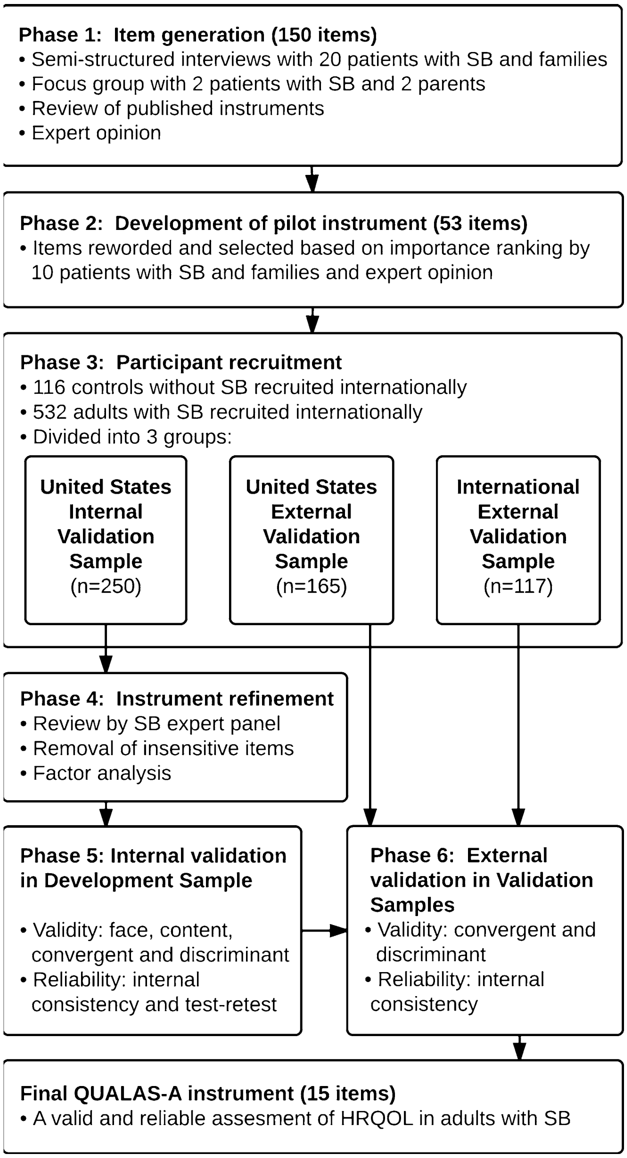

Fig. 1 Creation of the QUAlity of Life Assessment in Spina bifida in Adults (QUALAS-A). *SB* Spina bifida

The pilot QUALAS-A was self-administered. Questions were close-ended and numbered to avoid omission. Similar to several other instruments, questions reflected the last 4 weeks [12, 16, 17]. Responses used an evenly spaced 5-point response Likert scale (never, almost never, sometimes, almost always, always), an approach successfully used in other instruments [12, 13, 16–18]. Items focusing on a similar theme were grouped together to simulate the final QUALAS-A instrument.

Phase 3: Further participant recruitment

The pilot QUALAS-A and a demographic questionnaire were administered anonymously to a convenience sample of adults with SB and controls in an international cross-sectional survey (January 2013–September 2014). Adults with SB also completed the validated general World Health Organization Quality of Life instrument (WHOQOL-BREF) [6] and the International Consultation on Incontinence Questionnaire (ICIQ) [19]. In order to include adults from a variety of health care and social settings in the study, participants were recruited through local, national and international SB organizations via social media (see Acknowledgments) and at outpatient SB clinics at our center. Consent was either obtained online (computer based) or in person (clinic based). The QUALAS-A instrument was readministered 2 weeks later to participants recruited in clinic, to assess test–retest reliability. Healthy controls were recruited from among hospital staff and colleagues by email and completed QUALAS-A online. Participants did not receive incentive payments.

Phase 4: Refinement of instrument

A panel of SB experts reviewed responses for the 53 items and removed 3 items secondary to high missing rates (>50 %) and poor wording. To ensure sufficient statistical power to perform a factor analysis on 50 items, we would need approximately 250 participants (five participants/item) [20, 21]. We therefore randomly selected a sample of 250 participants from the United States (Internal Validation Sample). Only data from this Internal Validation Sample were used to develop the final QUALAS-A. In order to verify that our results were not spuriously arising from a restricted sample, we repeated the analysis in the entire study sample.

To further refine the instrument, we performed a principal axes method of factor analysis using varimax (orthogonal) rotation [22, 23]. The number of factors to extract was based on several criteria. (1) We examined the standard scree plot of the unrotated and rotated factor models, using the inflexion point after which the slope decreases linearly to indicate the optimal number of factors [22, 23]. (2) We extracted factor models with different numbers of factors and rotated them using five additional rotations (four orthogonal, one oblique) to ensure that the expected factor structure remained after the rotation. (3) All extracted factors needed at least 3 variables with loadings ≥ 0.4. (4) We examined the medical plausibility and clinical relevance of individual items loading on each factor. Based on these criteria, the final QUALAS-A instrument consisted of three domains of five items each. QUALAS-A is scored 0–100, where higher values signify higher HRQOL, and takes 5 min or less to complete.

Phase 5: Internal validation

We evaluated internal validity of the final QUALAS-A using the Internal Validation Sample described above.

Several types of validity were assessed to determine whether QUALAS-A measures what it is supposed to measure [24]. Face validity and content validity of the 15-question QUALAS-A were established through a review by patients, families and experts. Construct validity was additionally assessed through factor analysis. For each domain score, we calculated the mean, standard deviation (SD), median, range and the percentage of subjects scoring the minimum (floor) and maximum (ceiling). Reliability, or reproducibility, was assessed in several ways [24]. Internal consistency is a measure of reliability, referring to the degree of correlation between items in the instrument, and was measured by Cronbach's alpha (0.7–0.9 signifying good consistency without redundancy) [24]. Two-week test–retest reliability was assessed by intraclass correlation coefficient (ICC), which accounts for the direction of change, using a SAS macro written by Lu and Shara [25] ($\geq$0.7 indicating acceptable reliability).

Convergent validity evaluates the degree to which QUALAS-A scores converge with other instruments measuring similar outcomes. Conversely, divergent validity evaluates the degree to which QUALAS-A scores diverge from those measured by dissimilar instruments. Convergent validity and divergent validity assess redundancy or conceptual independence. We calculated Pearson correlation coefficients (r) among QUALAS-A domains and with WHOQOL-BREF domains. Criterion validity was assessed using Pearson correlation with the ICIQ. To further assess divergent validity, domain scores between the Internal Validation and Control Samples were compared using a t test. To further quantify effect size, we used a previously established method of dividing the mean difference between the Internal Validation and Control Samples by the SD of the control population [26, 27].

Several distribution-based approaches were used to determine the minimal clinically important difference that could be detected. Estimated point differences were determined using (1) 0.5 SD [28], (2) internal consistency or (3) test–retest reliability in the formula $\mathrm{SD} \times \sqrt{(1 - \text{reliability})}$ [29, 30]. We selected the most conservative, largest point difference calculated by the three methods as the minimally important score difference for each domain.

Phase 6: External validation

External validity was evaluated in the United States External Validation Sample ($n = 165$) and an exploratory International External Validation Sample ($n = 117$). Summary scores and internal consistency were calculated using methods similar to the Internal Validation Sample. Differences between Internal and External Validation Samples were compared using multivariate analysis of variance (MANOVA) to comprehensively account for all three QUALAS-A domains simultaneously. A critical $p = 0.05$ was used. All statistical analyses were performed using SAS software (version 9.4, SAS Institute, Cary, NC).

Results

Phases 1 and 2: Development of pilot instrument

The 53-item pilot instrument was developed in a multifaceted, patient-centered fashion (Fig. 1). Fifteen semi-structured interviews lasted a mean of 20 min. The focus group lasted 30 min.

Phase 3: Demographics

Of 614 online participants assessed for eligibility, 474 met inclusion criteria and participated (77.2 %) (Fig. 2). Ineligible participants tended to be 4 years younger than eligible participants ($p = 0.0002$). However, there was no significant difference between ineligible and eligible participants based on the gender or country of residence ($p \geq 0.79$). Of 95 eligible clinic participants, 58 consented to the study (61.1 %). Those refusing to participate tended to be 6 years older ($p < 0.0001$) but had a gender distribution similar to consenting participants ($p = 0.54$). Of 138 controls assessed for eligibility, 116 were included in the analysis (84.1 %). Ineligible controls were similar to eligible controls in terms of age, gender and country of residence ($p \geq 0.10$).

Median age of 532 participants was 32 years (32.7 % males, 85.0 % Caucasian), similar to 116 controls ($p \geq 0.08$) (Table 1). Participants lived with their parents (44.9 %) or spouse/partner (26.4 %), or alone (22.0 %). Half of participants were community ambulators (49.4 %) and 62.6 % had a ventriculoperitoneal shunt. Of the entire group, 22.0 % of participants were from outside the United States (46.3 % from outside North America and Europe, eTable 1). Participants from the United States lived in 44 states, encompassing 98 % of the country's population in 2011 (eTable 2) [31].

Phase 4: Factor analysis

After the removal of 3 items due to high missing rates and poor wording, 50 items were analyzed with factor analysis. A break in the slope of scree plots from two principal factor analyses, unrotated and varimax rotated, suggested the presence of 3–4 meaningful factors. The varimax-rotated model of 50 items confirmed 3 strong factors and 1 weak one, followed by a decreasing slope of minor factors (Table 2). Similar findings were obtained on the other 5

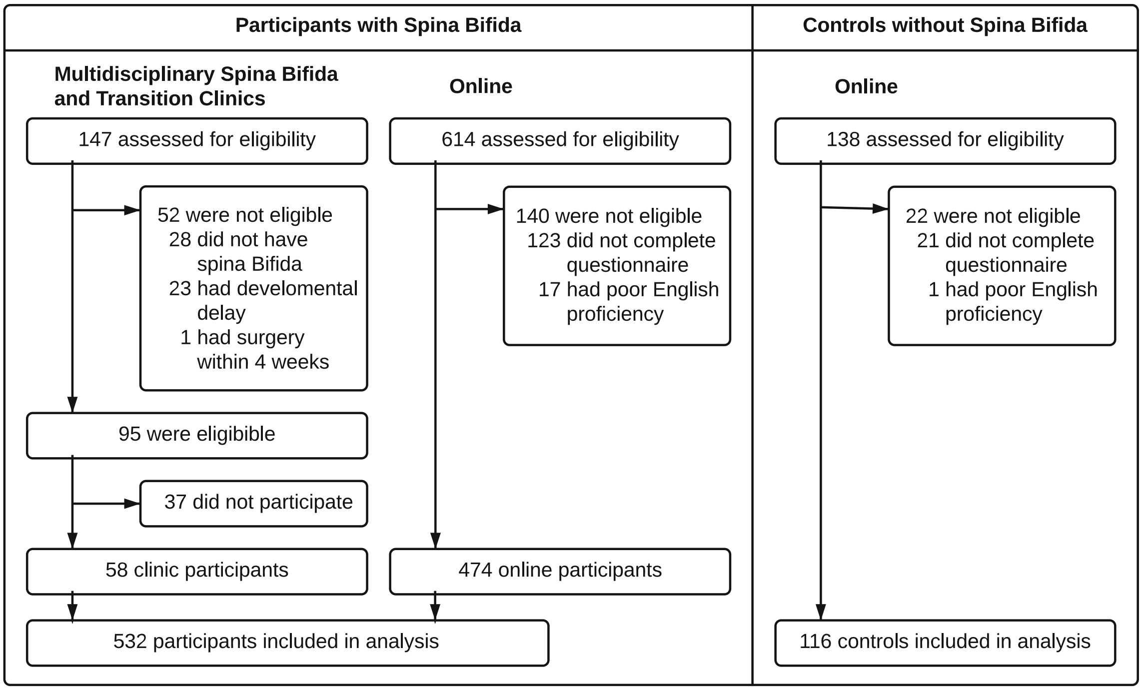

Fig. 2 Enrollment of adults into the study. Adults were enrolled at two clinics (Spina Bifida Clinic and the Transition Clinic) as well as online via social media. Enrollment commenced in January 2013 through September 2014 (20 months). Community controls, without spina bifida or children with spina bifida, were recruited from among hospital staff and colleagues

rotations. We extracted 3 factors and repeated the analysis to see whether the fourth could be ignored. The first 3 extracted factors were retained (Health and Relationships, Esteem and Sexuality, Bladder and Bowel). Factor 4 (Bathroom use) helped minimize cross-loadings between the first 3 factors, but was excluded from the final instrument because it was considered of little clinical importance and only 2 items had loadings ≥ 0.4. The 3 factors accounted for 63.6 % of the total variance.

Phase 5: Internal validation

A review by patients, families and experts established face validity and content validity of the final 15-question QUALAS-A. The Flesch Kincaid Grade Level test indicated a fifth-grade reading level. Characteristics of the QUALAS-A domain scores were evaluated, without evidence of floor or ceiling effects (Table 3a). Each of the three domains had robust internal consistency (Cronbach's alpha ≥ 0.70) and test–retest validity (ICC ≥ 0.77). The mean test–retest differences were small for each domain (range -5.0 to $+2$). Missing data were acceptably low for each domain (Health and Relationships 0.4 %, Esteem and Sexuality 12.8 %, Bladder and Bowel 0.4 %).

While an overall quality of life (QOL) item was not included in the final instrument because it was too broad, overall QOL was rated good to very good by 77.2 % of participants. Correlations between QUALAS-A domains were low ($r \leq 0.48$), indicating that QUALAS-A can differentiate between the 3 distinct HRQOL components (eTable 3). Correlations between QUALAS-A and WHOQOL-BREF domains were also low ($r \leq 0.60$), indicating that these instruments can be used concurrently. However, we noted high correlations with QUALAS-A Health and Relationship domain and WHOQOL-BREF ($0.63 \leq r \leq 0.71$). In addition, the Physical Health and Psychological domains of the WHOQOL-BREF were correlated with each other ($r = 0.74$) and assess some concepts similar to the QUALAS-A Health and Relationship domain. Of the 3 QUALAS-A domains, only Bladder and Bowel had a high correlation ($r = -0.70$) to the ICIQ, thus confirming criterion validity. The remaining QUALAS-A domains had low correlations with the ICIQ ($r \leq -0.33$).

QUALAS-A scores for controls were significantly higher than those in the Internal Validation Sample for all 3 domains ($p < 0.0001$), each with a large effect size (≥ 0.9) (eTable 4). Similar to the Internal Validation Sample, missing data were low for each domain (Health and Relationships 5.2 %, Esteem and Sexuality 15.5 %, Bladder and Bowel 0.0 %).

Using several distribution-based approaches, we calculated that the minimally important difference for the Health

Table 1 Population characteristics

Patient characteristic	Participants ($n = 532$)	Controls ($n = 116$)	*p* value
Age, mean (SD), years	32.1 (11.0)	34.0 (10.5)	0.09
Male gender, no. (%)	174 (32.7 %)	48 (41.4 %)	0.08
Race			
Caucasian	452 (85.0 %)	100 (86.2 %)	0.89
Hispanic	26 (4.9 %)	2 (1.7 %)	
African-American/African	15 (2.8 %)	1 (0.9 %)	
African-Caribbean	1 (0.2 %)	0 (0.0 %)	
Southeast Asian	5 (0.9 %)	2 (1.7 %)	
East Asian	7 (1.3 %)	4 (3.5 %)	
Arab	1 (0.2 %)	3 (2.6 %)	
Native[a]	8 (1.5 %)	1 (0.9 %)	
Multiethnic	17 (3.2 %)	3 (2.6 %)	
Household arrangements, no. (%)			
Parent(s)	239 (44.9 %)	18 (15.5 %)	<0.001
Spouse/partner	140 (26.3 %)	71 (61.2 %)	
Alone	117 (22.0 %)	16 (13.8 %)	
Housemate(s)	12 (2.3 %)	16 (13.8 %)	
Grandparent(s) only	4 (0.8 %)	0 (0.0 %)	
Uncle/aunt only	5 (0.9 %)	0 (0.0 %)	
Caregiver only	7 (1.3 %)	0 (0.0 %)	
Siblings only	5 (0.9 %)	0 (0.0 %)	
Unknown	3 (0.6 %)	0 (0.0 %)	
Mobility, no. (%)			
Community ambulator	263 (49.4 %)	115 (99.1 %)	<0.001
Not a community ambulatory	264 (49.6 %)	1 (0.9 %)	
Unknown	5 (0.9 %)	0 (0.0 %)	
Ventriculoperitoneal shunt, no. (%)			
Yes	333 (62.6 %)	0 (0.0 %)	<0.001
No	170 (32.0 %)	116 (100.0 %)	
Unknown	29 (5.5 %)	0 (0.0 %)	
Clean intermittent catheterizations, no. (%)			
Yes	398 (74.8 %)	1 (0.9 %)	<0.001
No	134 (25.2 %)	115 (99.1 %)	
Social urinary continence (≥4 h dry), no. (%)			
Yes	385 (72.4 %)	113 (97.4 %)	<0.001
No	141 (26.5 %)	2 (1.7 %)	
Unknown	6 (1.1 %)	1 (0.9 %)	

Totals may not add up to 100 % due to rounding

SD standard deviation

[a] Native American, Alaska Native, Inuit, Aboriginal, Maori, etc.

and Relationship domain was ≥10 (range of values 8.2–10.0). The minimally important difference for the Esteem and Sexuality and Bladder and Bowel domains was ≥15 (range of values 9.8–13.4 and 12.2–13.0, respectively).

Results of the analysis on the entire sample ($n = 532$) were not significantly different from the analysis on the Internal Validation Sample ($n = 250$) (data not shown).

Phase 6: External validation

Characteristics of the QUALAS-A domain scores in the two External Validation Samples were evaluated without evidence of floor or ceiling effects (Table 3b, c). QUALAS-A had good statistical properties in the United States and exploratory International External Validation Samples

Table 2 Factor analysis of responses from 250 adults with spina bifida living in the United States

Item	Factor 1	Factor 2	Factor 3	Factor 4
Able to do fun things	68*	20	9	13
Time with friends	65*	−1	9	16
Close friendships outside family	64*	13	15	3
Overall health	56*	35	22	−4
People saw you for more than health problems	46*	33	11	24
Bother by others helping	16	60*	13	24
Having future children	−13	56*	10	−12
Embarrassment about your look	28	55*	24	−1
Bother by sexual in/activity	26	51*	22	−18
Future sexual satisfaction impacted by health	27	51*	32	−15
Worry about pads being noticed	13	28	70*	11
Bother by bowel leak	5	11	59*	7
Bother by urine leak	28	15	54*	1
Urine problems stop you from fun things	40	30	48*	−7
Bother by waiting for bowel movement	14	19	48*	9
Easy to use toilet at home	5	7	6	71*
Easy to use bathroom by yourself	13	0	2	66*

Of the 4 factors analyzed, the first 3 extracted factors were retained (Health and Relationships, Esteem and Sexuality, Bladder and Bowel). Factor 4 (Bathroom use) was not included in the final instrument because it was the least clinically useful and had only 2 items with loadings >0.4. A similar analysis was performed on 35 other items assessed (data not shown). Factor loadings in the table are multiplied by 100 and rounded to the nearest integer

* Factor loading value is greater than the geometric mean (root mean square) of all values in the matrix

(Cronbach's alpha ≥ 0.68 and ≥ 0.71, respectively). No significant difference in QUALAS-A scores was noted between three Validation Samples overall ($p = 0.55$), between the two United States samples ($p = 0.37$) or the Internal Validation and International Samples ($p = 0.34$).

Discussion

We present a novel HRQOL instrument developed specifically for adults with SB, which has been validated in United States and International population samples. QUALAS-A is composed of items relevant to adults with SB, their families and clinicians, including comprehensive domains focusing on Bladder and Bowel function, as well as sexuality. Questions regarding sexuality are particularly important, since they are rarely discussed with health professionals [32] and individuals with SB are often socially infantilized [33]. Finally, we report domain-specific minimally important differences in scores, which may help patients, clinicians and researchers to differentiate potential clinically important differences from random measurement errors.

Three previously published SB-specific HRQOL instruments have serious limitations. The Fecal Incontinence and Constipation tool and the Hydrocephalus Outcome Questionnaire were validated only for parents of children under 18 years old [12, 34]. In addition, the Hydrocephalus Outcome Questionnaire was developed without input from people with SB. On the other hand, the HRQOL-SB instrument was the only one developed with input from adults with SB and to have a patient-administered version [13]. Furthermore, each instrument has at least 44 items, making it cumbersome and time-consuming to administer. Since none of the 3 instruments was designed using factor analysis, all remain suspect for item redundancy and appropriate domain groupings. Since their initial publication, none of these instruments have been externally validated. Also, when scoring these instruments, it remains unclear what score difference, or change, is clinically relevant. Finally, aside from the Fecal Incontinence and Constipation tool, these instruments do not include items that deal specifically with bladder or bowel care or incontinence, areas demonstrated to be important to all patients and families in our focus group and interviews.

Rather than being a simple binary variable (good/bad), HRQOL is a continuous, multidimensional and subjective concept affected by multiple clinical and psychosocial variables. Our findings indicate that most adults with SB describe themselves as having good to very good overall QOL even when burdened with significant physical limitations, which supports similar findings in adolescents with SB [35]. This finding suggests that adults with SB

Table 3 Domain characteristics of the self-reported QUALAS-A. Statistical properties were analyzed in (a) the Internal Validation Sample, (b) the United States External Validation Sample and (c) the International External Validation Sample

Domain	*N*	% missing	Mean (SD)	Median (range)	% scoring minimum	% scoring maximum	Cronbach's alpha	Test–retest reliability (ICC)
a. Internal Validation Sample (*n* = 250)								
Health and Relationships	249	0.4	67.8 (20.0)	70.0 (0.0–100.0)	0.4	6.8	0.79	0.83
Esteem and Sexuality	218	12.8	57.8 (24.4)	60.0 (0.0–100.0)	1.8	3.7	0.70	0.84
Bladder and Bowel	250	0.0	61.6 (25.5)	60.0 (0.0–100.0)	0.4	8.0	0.74	0.77
b. United States External Validation Sample (*n* = 165)								
Health and Relationships	165	0.0	67.5 (18.9)	70.0 (10.0–100.0)	0.0	2.4	0.75	n/a
Esteem and Sexuality	134	18.8	57.8 (24.0)	55.0 (0.0–100.0)	2.3	3.8	0.68	n/a
Bladder and Bowel	165	0.0	63.7 (25.5)	65.0 (10.0–100.0)	2.4	12.1	0.75	n/a
c. International External Validation Sample (*n* = 117)								
Health and Relationships	113	3.4	67.4 (18.7)	68.8 (25.0–100.0)	0.0	2.7	0.74	n/a
Esteem and Sexuality	98	16.2	53.7 (25.3)	55.0 (0.0–100.0)	1.0	4.1	0.77	n/a
Bladder and Bowel	113	3.4	62.2 (25.6)	65.0 (0.0–100.0)	2.7	9.7	0.71	n/a

QUALAS-A QUAlity of Life Assessment in Spina bifida for adults, *SD* standard deviation, *ICC* intraclass correlation coefficient

may have a response shift [36], a process in which individuals with chronic conditions change their priority for different components of HRQOL or have an altogether different idea about HRQOL. The very concept of HRQOL may develop differently in people with and without SB, resulting in a cutoff for a poor QOL being significantly lower than for those without SB. For this reason, QUALAS-A scores of healthy controls may hold no clinical relevance and be solely of statistical value in the validation process. In addition, rather than treating QUALAS-A scores as absolute constants, scores may be best compared longitudinally to the same person over time or between individuals with the same or similar chronic conditions. Finally, QUALAS-A evaluates the impact of SB on HRQOL in adults. For that reason, it should be used in conjunction with a generic HRQOL measure if one wishes to assess overall QOL.

Our study had a robust design, resulting in a validation across several large, heterogeneous populations. The levels of missing data for the QUALAS-A were low across all domains. This was even true for the Sexuality and Esteem domain, which contains sensitive items that some adults are reluctant to answer, whether or not they have SB. Similar to other instruments, when 1 of the 5 responses is missing, we suggest considering the remaining items in the domain [6].

Our study had several limitations. Participants were a convenience sample and may not be representative of the general SB population. Participants were highly motivated, since they completed the 20-min survey without compensation, and possibly had fewer developmental and functional limitations than general SB population. Selection bias is unlikely to play a large role, as gender and geographical distributions were similar between participants and those who either refused to participate or were ineligible. Although older patients were more likely to refuse to participate in clinic, it was precisely the older patients who were more likely to enroll online. In addition, it is unlikely that a significant number of adults without SB participated in the study as presumed adults with SB due to focused advertising through SB organizations and the considerable time investment to complete the study. While we did not use anchor-based methods to calculate minimally important differences, distribution- and anchor-based methods have been shown to give comparable results [37]. Being a validation study, we did not assess factors associated with higher HRQOL. This will require further investigation. Finally, although we validated the English version of QUALAS-A in an exploratory International Sample, use in a wider variety of settings will require a cross-validation in other countries and into other languages.

Conclusion

QUALAS-A is a short, valid tool for evaluating HRQOL in adults with SB. It assesses a comprehensive array of domains, making it a useful instrument in both clinical and research settings.

Acknowledgments We would like to thank the hundreds of individuals with spina bifida and their parents and caregivers participating in this study. We also want to thank the following individuals and groups for their help: *Concept and Content Consultant:* Kathleen J. Sawin, PhD CPNP-PC FAAN (College of Nursing, University of Wisconsin-Milwaukee/Children's Hospital of Wisconsin); *Content Consultants:* Pediatric Urology: John Paul Capolicchio, MD (Montreal Children's Hospital, McGill University), Mohamed Elsherbini, MD (Montreal Children's Hospital, McGill University), Walid Farhat, MD (Hospital for Sick Children, University of Toronto), Richard Grady, MD (Seattle Children's Hospital, University of Washington), Roman Jednak, MD (Montreal Children's Hospital, McGill University), David B. Joseph, MD (University of Alabama at Birmingham), Alison C. Keenan, MD (Riley Hospital for Children, Indiana University Health), Martin A. Koyle, MD (Hospital for Sick Children, University of Toronto), Andrew L. MacNeilly, MD (British Columbia Children's Hospital, University of British Columbia), JL Pippi Salle, MD (Hospital for Sick Children, University of Toronto), Melissa A. Young, CPNP (Riley Hospital for Children, Indiana University Health). Pediatrics: Timothy J. Brei, MD (Seattle Children's Hospital, University of Washington), Joseph O'Neil, MD (Riley Hospital for Children, Indiana University Health). Clinical Pediatric Psychology: Khush Amaria, PhD (Hospital for Sick Children, University of Toronto). Health Literacy: Cynthia Latty, RN (Riley Hospital for Children, Indiana University Health); *Statistics Consultants:* Robert Haley, MD (University of Texas Southwestern), Patrick Monahan, PhD (Richard M. Fairbanks School of Public Health, Indiana University); *Participant Recruitment:* Robin Bowman, MD (Lurie Children's Hospital of Chicago, Northwestern University), Timothy J. Brei, MD (Seattle Children's Hospital, Seattle Children's Hospital), Jerry Clayton, MD (Children's Hospital Colorado, University of Colorado), Dominic C. Frimberger, MD (The Children's Hospital of Oklahoma, University of Oklahoma), Betsy Hopson, MSHA (Children's Hospital of Alabama, University of Alabama at Birmingham), Jill Mazurek, MD (Indiana University Health), Brandon G. Rocque, MD (Children's Hospital of Alabama, University of Alabama at Birmingham), Dorota A. Szczepaniak, MD (Riley Hospital for Children, Indiana University Health); *Record Entry and Review:* Sable Amstutz, MD (Indiana University Health), Kyle Hardacker (Indiana University Health), Zoe King (Indiana University Health), Sophie Mazurek (Indiana University Health), Meredith Metcalf, MD (University of Tennessee Health Sciences Centre), David Yang, BSc (Indiana University Health); *Organizations assisting in participant recruitment:* International: International Federation for Spina Bifida and Hydrocephalus. United States: Spina Bifida Association, Spina Bifida Associations of Alabama, the Carolinas, Community of Memphis, Connecticut, Georgia, Greater New England, Illinois, Indiana, Kentucky, Nassau County, New Orleans, North Texas, Texas, Washington, Western Pennsylvania, Wisconsin, SB Resource Network, Spina Bifida Connection, National Birth Defects Prevention Network, Disabled Children of Vietnam Veterans. Canada: Spina Bifida & Hydrocephalus Association of Canada, Spina Bifida & Hydrocephalus Association of Ontario, Association de Spina-Bifida et d'Hydrocéphalie du Québec. Europe: Spina Bifida Hydrocephalus Ireland (Ireland), Mid-West Spina Bifida and Hydrocephalus Association (Ireland), Cork Spina Bifida and Hydrocephalus Association (Ireland), Spina bifida Hydrocephalus Information Networking Equality (SHINE, UK), Arbeitsgemeinschaft Spina Bifida und Hydrocephalus e. V. (ASBH, Germany), Swedish National Association for Disabled Children and Young People (FUB) (Sweden), Asociación Bizkaia Elkartea Espina Bífida e Hidrocefalia (Spain), Fundacja Spina (Poland), Spina Bifida ir Hidrocefalija asociacija (SBHA) (Lithuania). Australia and Oceania: Spina Bifida Foundation Victoria (Australia), Spina Bifida Hydrocephalus Queensland (Australia), Spina Bifida Awareness (New Zealand). Middle East: Lebanese Association for Neuromuscular Diseases for Hope (Lebanon). South America: Associação de Espinha Bífida e Hidrocefalia (AEBH, Brazil). Africa: Festus Fajemilo Foundation (Nigeria), National Council for Persons with Physical Disabilities in South Africa (South Africa).

Conflict of interest Dr. Misseri is a site Investigator and Shelly King is a site Coordinator of the Allergan study 191622-120. None of the other authors declare potential competing personal interests. No authors declare any potential financial conflict of interest.

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